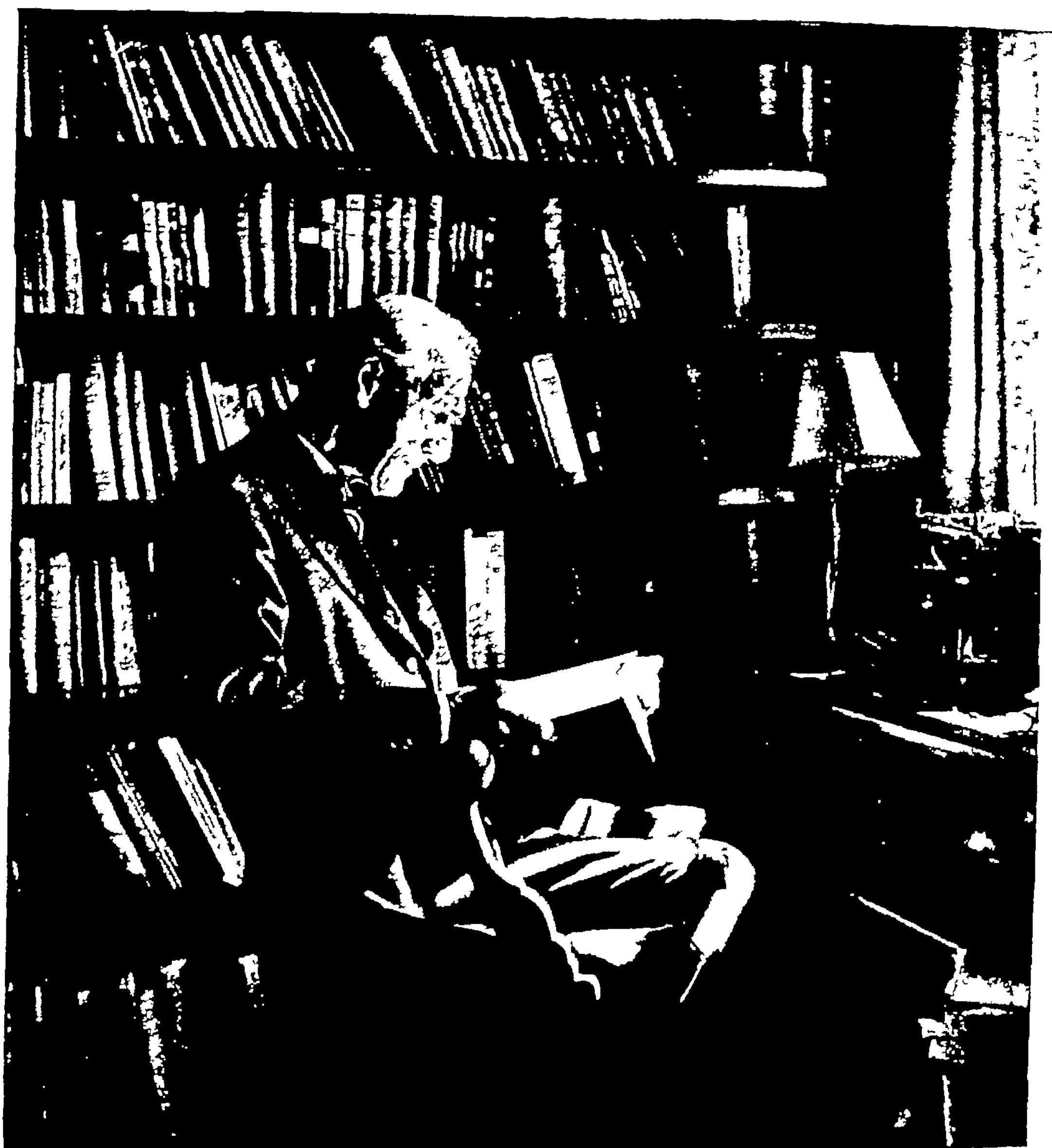


Sir Arthur Keith

The Anatomy of Congenital Pulmonary Stenosis

Sir Russell Brock
M.S F.R.C.S F.A.C.S (Hon.)
Thoracic Surgeon to Guy's Hospital
Surgeon to the Brompton Hospital



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INTRODUCTION

THIS account of the anatomy of congenital pulmonary stenosis was written in its greater part over five years ago at the time when great interest had recently been aroused in the condition and many cardiologists, surgeons, radiologists and others were engaged in the investigation and treatment of cases of pulmonary stenosis Its publication was, however, deliberately delayed because I realised that with a steadily increasing experience in the management and surgical treatment of these cases I was acquiring much additional information. Although, in the meantime, a number of accounts of the anatomy of pulmonary stenosis have appeared, I have not regretted this delay for the fuller experience has been of greater value

The term pulmonary stenosis is itself, of course, not precise, in this book it is used as a comprehensive term covering all varieties of obstruction to the blood flow to the lungs from congenital malformation. The individual varieties of pulmonary stenosis will be indicated by their appropriate names.

My interest in the development and surgical anatomy of the condition arose from two reasons. First, it is more satisfactory to know why a thing occurs and it seemed obvious that there must be an explanation for the common occurrence of such a peculiarly constant form of maldevelopment as pulmonary stenosis, even though its varieties are several. Second, my interest in the relief of pulmonary stenosis by direct operation made it essential to understand the anatomical formation both of the stenosis itself and of the muscular structure of the ventricular walls in its neighbourhood. As a corollary to this it is necessary to emphasise the need for careful and complete study of the normal morbid anatomy of a disease process when it is being investigated and treated. This remark may be superfluous but the fact remains that many people are handling cases of pulmonary stenosis, including performing operations upon them, without any more than a second-hand and often rather hazy picture of the anatomical condition present. This is quite apart from the practice of making an empiric diagnosis of pulmonary stenosis and advising and performing a stereotyped operation for its indirect relief without attempting, at operation, to make the diagnosis of the particular variety as exact as possible. The information generally available about the anatomy of the condition is, moreover commonly accepted without any personal verification or study This is obvious from the popular conception of the nature of the obstruction, especially the infundibular form.

In any case the introduction of such an important step as operation for the relief of pulmonary stenosis demands a reinvestigation of the whole subject and verification of past observations and descriptions What might have been acceptable as a general description for a condition which was of little interest other than as an untreatable clinical state, or as a range of congenital malformations, can be inadequate if exact surgical attack is to be based on it It is imperative in such new circumstances to re-open the whole problem and to confirm, amplify or modify the older descriptions and conceptions

It is also important to realise that there may be a great difference between the anatomical state of affairs as observed after death in a specimen in a condition of muscular contraction aggravated by fixation and hardening, and the state that exists during life in an active, functioning muscular structure such as the heart The surgeon who observes at operation the condition both outside and inside the living heart will see definite and significant differences in the same specimen after death We must remember this difference between the anatomy of the living heart and the dead heart and that, in so far as this feature is concerned, post-mortem examination may be deficient and misleading instead of being the acme of truthful observation it is usually thought to be.

The ordinary conception of the right ventricle is often that of a rather simple muscular structure which expands to receive blood from the atrium and then contracts to expel it In fact the arrangement is much more complex than this, as will appear from the succeeding description The formation, structure and function of the outflow portion of the right ventricle in particular is quite complex and is of great practical importance in relation to the stenoses that affect it

The general paucity of knowledge about the anatomy of pulmonary stenosis at the time of the introduction of surgical treatment was surprising because reference to the literature reveals a whole wealth of first-class information that is interesting, informative and even enthralling Much of this has remained neglected or forgotten and to the surgeon seeking practical information it is rather like digging and finding gold In this connection one must mention especially the writings of Sir Arthur Keith which, as always, are charmingly and simply written and full of scholarship, clear observation and deduction No one interested in the subject should fail to read the series of lectures published in the *Lancet* between 1904 and 1924 As long ago as 1904 Keith wrote in the introductory paragraph to his Hunterian Lecture at the Royal College of Surgeons on *The evolution and action of certain muscular structures of the heart* 'It is with a feeling of keen disappointment that men who seek to extend our knowledge of the origin and development of the human heart lay down any of the standard treatises on the function and diseases of that organ Their labour as far as one may judge from the perusal of such works lies outside the field of practical medicine and is useless to physician and physiologist alike'

Keith had to wait nearly half a century to see his work on the nature and development of pulmonary stenosis bear fruit, but it is now possible to say that his observations and explanations have been of immense help in planning and

developing a logical surgical approach to the direct operative relief of pulmonary stenosis

I have to thank Professor T B Johnston for reading through my manuscript, and making numerous valuable suggestions and correcting several errors

My secretary Miss Chris Jones, has spent many tiring hours on the preparation and scrutiny of manuscript and proofs and I am very grateful to her

Sir Arthur Keith gave permission to use certain of his diagrams and also selected the frontispiece from among his several photographs, it was taken by Mr John Miller of Barnet Messrs Edward Arnold and Company have given permission to reproduce Figures 1 and 6 from *Human Embryology and Morphology*

Messrs Baillière, Tindall, and Cox have given permission to reproduce Figure 9

I am very grateful to Dr Prinzmetal for his kindness in sending me a short strip from one of his splendid films which demonstrates the peristaltic-like movements of the infundibulum.

I am also grateful to the various colleagues who have given indispensable aid by recording electromanometric pressures both at cardiac catheterisation and at operation. Among these are Dr D C. Deuchar, Dr R. J Shephard, Dr R. Gibson, Dr P Fleming and Dr L. Brotmacher Dr Maurice Campbell has kindly allowed me to use Figure 34 from one of his articles Figure 51 was first published in the *British Heart Journal*

I acknowledge with gratitude the help so readily and skilfully given with the preparation of many of the photographs, drawings and pressure records by the Members of the Department of Medical Illustration of Guy's Hospital and of the Photographic Department of the Brompton Hospital.

CHAPTER I

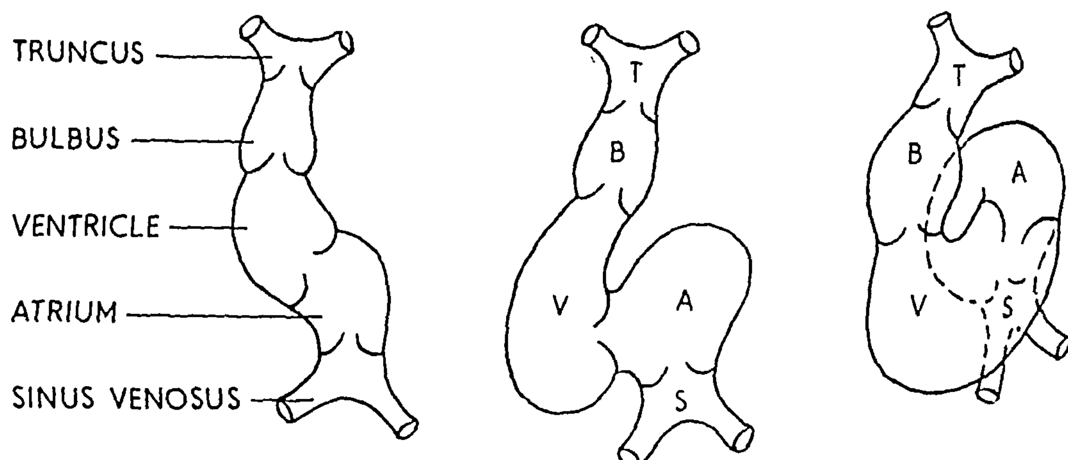
The comparative anatomy and embryology of pulmonary stenosis

It is easier to understand the embryology of pulmonary stenosis from a simple knowledge of the comparative anatomy The complicated developmental features of the human heart are difficult to follow until it is appreciated that in its development the heart passes through most of the stages of formation of the heart of the lower orders of animals. In fact most of the congenital malformations represent the heart, fixed as it were, at a more primitive stage of development: one that can be recognised as belonging to that commonly seen in certain of the lower orders, with the addition of adaptations resulting from the arrested development.

Maude Abbott (1936) in her classical *Atlas of Congenital Cardiac Disease* remarks: 'The most bizarre combinations of defects can usually be interpreted quite simply, as due to early arrest of development, marked it may be by ingenious structural adaptation of growth', and 'Much light is cast upon the development of the mammalian heart, and incidentally, upon the period at which arrest of growth has taken place in cardiac anomalies of the graver sort by a comparative study of the adult fish, amphibian and reptilian organ. The truly extraordinary way in which these various orders in the ascending vertebrate scale mirror the successive stages through which the human heart passes in very early intrauterine life is one of nature's most spectacular and impressive feats, presenting as it does a complete review of this organ's evolution down to the closure of the cardiac septa in the eighth week of foetal life.'

The following account does not pretend to be original; the description is entirely second-hand although illustrated by a number of comparative anatomical specimens personally collected and studied. It is not possible for a practising surgeon to survey and verify the whole of this vast comparative and developmental problem, it is essential to rely upon the work of others. The examination, dissection and description of the human material is, however, the result of personal work.

The primitive tubular heart consists essentially of five parts, the sinus venosus, the atria, the ventricles, the bulbus cordis and the truncus arteriosus (Fig. 1). It



1 Diagrams to show the primitive tubular heart of five chambers and how, as the heart lengthens and becomes folded on itself, the bulbus comes to lie beside the atrium and ventricle (*After Keith, 'Human Embryology and Morphology', Arnold*)

can be said at once that what concerns us most in the present problem is the formation and fate of the bulbus cordis. Thus Keith (1909) states 'one of the greatest discoveries since Peacock's time is now only dawning, but every year increases our assurances of its truth—viz that there is a fourth part or chamber in the mammalian heart which hitherto we have taken no cognisance of . . The fourth part is the bulbus cordis . . We have good reason for believing that, in the same manner as the sinus venosus has become incorporated in the right auricle, the bulbus has become included in the right ventricle, forming the part loosely termed its infundibulum' Although Keith mentions only four parts of the primitive heart, later (1933) he includes the truncus arteriosus as a fifth element and it is really more correct to say that the bulbus is absorbed partly into the ventricles and partly into the truncus, of which it forms the pericardial part. He goes on to say, 'The credit of this discovery belongs to Alfred Greil, prosector in the University of Innsbruck. He traced the fate of the bulbus by a prolonged study of the hearts of developing vertebrates (Greil 1903). Independently of him I had reached the same conclusion from an investigation of malformed human hearts and of the hearts of vertebrate animals. A large number of the very commonest malformations of the human heart are due to an arrest of the process

which ends in the incorporation of the bulbus cordis in the right ventricle. The great majority of cases of congenital stenosis of the pulmonary artery are of this nature.

The bulbus cordis is best shown in fishes, i.e. gill breathers, Figures 2–4 show examples in the skate, shark and angler fish and demonstrate that it varies much in size and shape in the different classes Quain (1929) makes some interesting comments on its nomenclature, about which there is sometimes confusion. The names that have been used for it are *conus arteriosus*, *bulbus arteriosus*, *truncus arteriosus* and *bulbus cordis*

Walmesley in *Quain's Anatomy* pointed out that, *conus arteriosus* is now used in human anatomy for a definite part of the right ventricle; *truncus arteriosus* is the name now usually applied in human embryology to that part of the common ventral vessel from which the pulmonary aorta and the proximal part of the systemic aorta are derived and the structure of whose wall, from the beginning, is that of a vessel. In the Teleostei this section of the ventral vessel is enlarged and its wall is thickened by a great development of smooth muscle and elastic tissue fibres, but it is entirely free from cardiac muscle; the name *bulbus arteriosus* is then used for it.

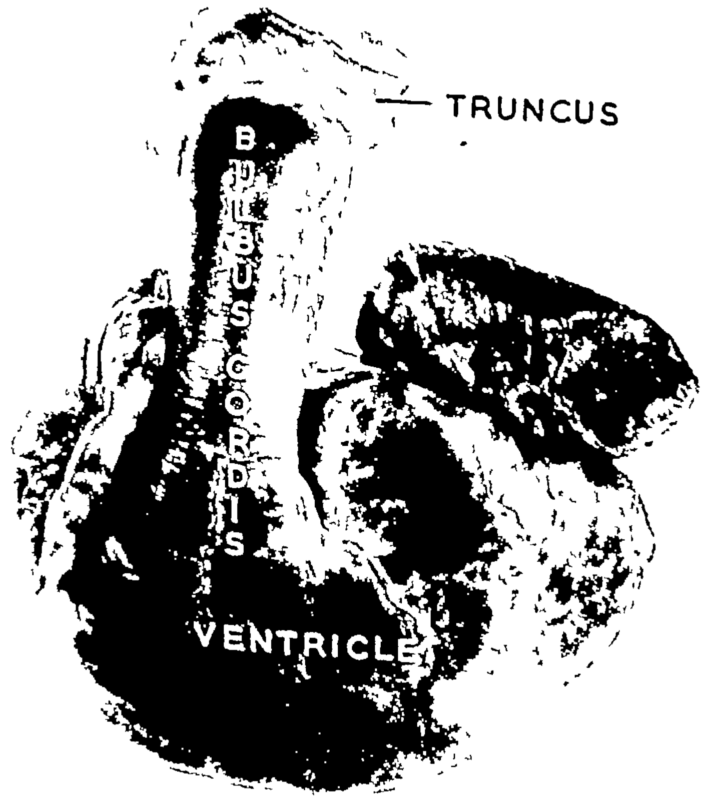
'The name *bulbus cordis* is thus the most suitable to be applied to that region which is the most distal part of the heart and which yet forms the most proximal part of the exit tube from the heart and it is to be considered a cardiac chamber for in its interior it carries the bulbar valves and in its wall there is cardiac muscle. It is present in all vertebrate embryos but in most forms it undergoes an ontological retrogression through the absorption of its proximal part into the ventricle and of its distal part into the truncus arteriosus, it remains persistent as a clearly separable chamber of the heart only in the Elasmobranchii Ganoids, Dipnoi and amphibia.

According to Waterston (1918) Langer first suggested the name bulbus cordis and it was then generally recognised and adopted

The specimens depicted in Figures 2–4 clearly show the muscle in the wall of the bulbus cordis especially Figure 3 (the heart of a basking shark) in which the muscle is darker The triple row of bulbar valves is also seen. The apparent constriction where the bulbus joins the ventricle should be noted (Fig. 2*b*) in view of what will be said later about constriction at the bulboventricular junction in cases of arrested fusion of the bulbus in man. The bulboventricular spur should also be noted

In contrast to the large size of the bulbus cordis in the fish heart there is no separate bulbus cordis in reptiles as is shown in the turtle's heart in Figure 5

It is necessary to consider the reptilian heart because of other striking features which it presents The turtle's heart shown in Figure 5 has two atria, the right being much larger than the left, and one common ventricle. The most striking feature is that three large vessels leave the common ventricle; a pulmonary artery and two systemic aortae, a right which supplies the abdominal viscera and the lower extremities and a left which supplies the head and the upper extremities.

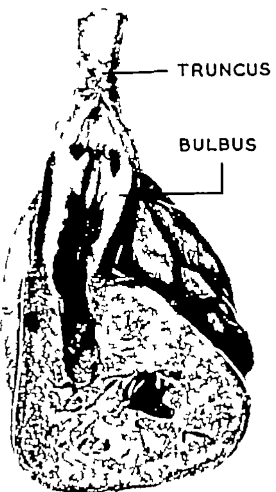


a

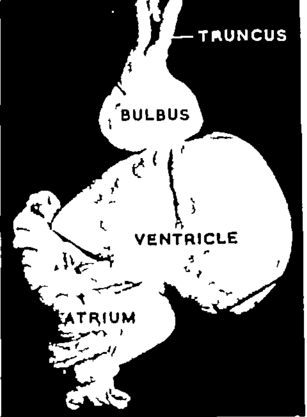


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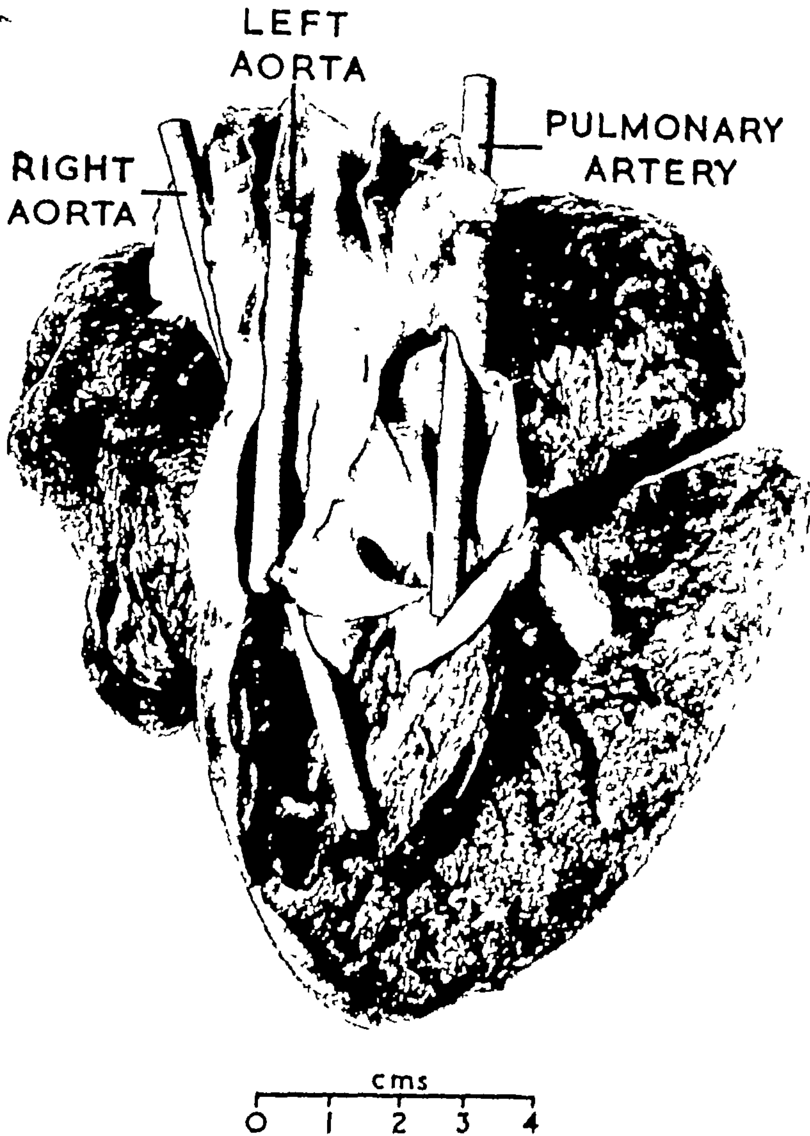
The heart of a skate to show the common ventricle and bulbus cordis Note the bulboventricular spur



3 The heart of a basking shark to show the common ventricle the bulbus cordis and the truncus arteriosus The continuity of ventricular and bulbar muscle is well shown.



4 The heart of an angler fish (Museum of Royal College of Surgeons) The bulbus forms a solid almost globular mass The atrium and ventricle are well seen.



5 The heart of a turtle showing the two aortae and the pulmonary artery, also the muscular septa or trabeculae within the single ventricle

The three vessels each have bicuspid semilunar valves and as they arise from the heart are intimately connected in a firm fascial sheath As they pursue their course they undergo a clockwise rotation from right to left reminiscent of the spiral arrangement of the embryonic swelling in the primitive truncus arteriosus

Although the ventricle is single its chamber is by no means simple It is occupied by a fine sponge-like mesh and in addition is partly divided by a bulbo-ventricular ridge into a smaller ventral chamber from which the pulmonary artery arises and a larger dorsal chamber The right systemic aorta arises from the extreme upper right-hand corner of this and just in front arises the left aorta The dorsal part of the ventricle is itself incompletely divided into shallow right and left cavities by an obliquely placed septum derived from the loose muscular trabeculae of its lower wall That on the right side, known as the cavum venosum,

receives the venous blood from the right atrium while that on the left, the cavum arteriosum, receives the oxygenated blood from the left atrium but gives off no vessel (Abbott, 1936)

Abbott states that these three arterial trunks are present together for a very short time in the human embryo, but this seems incorrect. On the other hand they provide a phylogenetic basis for Spitzer's theory of the mechanism of transposition of the great vessels In the human heart the reptilian aorta is obliterated by the normal clockwise rotation of the primitive cardiac tube. Spitzer believes that various forms of transposition arise from absent or incomplete fusion. In transposition the aorta is not a transposed aorta but is a reopened right aorta, the left having been suppressed.

Spitzer also points out that the phylogenetic presence of a right aorta is reflected in one part of the configuration of the normal right ventricle; this is the small fossa lying between the *crista supraventricularis* and the anterior cusp of the tricuspid valve and which really represents the outflow tract of the reptilian right aorta.

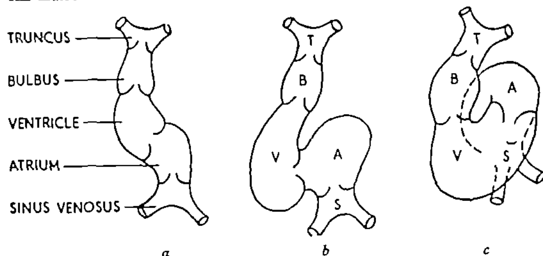
CHAPTER II

The developing human heart

In describing the embryology of the human heart relevant to pulmonary stenosis it is desirable to concentrate chiefly on the outflow side to avoid making the description too complex It is not possible to adhere to this entirely since in the course of the development of the human heart the venous and arterial sides are brought close together The formation of the right atrio-ventricular orifice, for instance, is intimately concerned with closure of the ventricular septum and the development of those parts of the right ventricle that concern us, and with the disappearance of the bulboventricular spur

The primitive tubular heart (Fig 6*a*) undergoes irregular growth so that it becomes folded on itself to give the shape of a simple, spiralled S as shown in Figures 6*b* and 6*c* in which the primitive atria and the primitive ventricle are seen with the bulbus cordis and the truncus arteriosus, the bulboventricular groove should be noted From this primitive outline the final form of the heart is reached The parts about which we need to concern ourselves in this account are

(i) The formation of the two ventricles
(ii) The division of the truncus arteriosus into the aorta and pulmonary artery
(iii) The formation of the right atrioventricular orifice
(iv) The closure of the ventricular septum
(v) The incorporation of the bulbus cordis into the right ventricle



6 Primitive tubular heart with formation of primary chambers (*After Keith*)

(i) *The formation of the two ventricles*

According to Keith (1909c) a bifid condition of the apex of the heart is not uncommon as an abnormality in man and is a normal form in some mammals such as the dugong Figure 7 shows a bifid apex in the heart of a man who had pulmonary valvar stenosis with a persistent foramen ovale. Keith states it was difficult to give a satisfactory explanation when the ventricular septum was regarded as an upgrowth from the floor of the primitive ventricle but, The opposite is the case; the ventricular cavities are downgrowths or evaginations of the primitive cardiac tube, the septum being left between them during development. The upper margin of the septum thus represents a part of the lumen of the primitive heart; in the upper margin is developed the auriculo-ventricular bundle.'

Baxter (1953) writes. 'This septum is really little more than the original floor of the cavity persisting at its original level, the two subdivisions of the cavity enlarging downwards on each side of it The ventricles have become deeper and the septum proportionately higher, but that the septum has *not grown upward* is evident from the fact that it retains its old level of attachment to the lower atrioventricular cushion this level is *original and persistent throughout*

Keith, after mentioning the auriculo-ventricular bundle developing in the upper margin of the septum goes on to say, In malformed hearts the bundle has a definite relationship to the interventricular foramen. On the right septal wall the bundle begins in the node situated behind the tendon of the septum and passes forwards on the lower margin of the foramen.

The exact relationship of the atrioventricular bundle to the septal defect and to the other structures in the right ventricular outflow tract (*infundibulum*) is obviously of great practical importance when intracardiac surgery is being planned

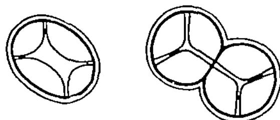


7 Human heart with bifid apex

(ii) The division of the truncus arteriosus into the aorta and the pulmonary art

The truncus arteriosus, the single vessel that conveys blood from the pri tive heart, is divided into its two final components by the development of t prominent ridge-like thickenings of its endocardial lining, these ridges, the ri and left spiral ridges, fuse to form the aortopulmonary septum which divides unabsorbed part of the bulbus into an aortic and a pulmonary trunk Below t spiral septum four endocardial cushions form in the distal part of the bulb cordis near its junction with the truncus, the right and left cushions fuse togeth to form a septum (the distal bulbar septum) with which the spiral septum fus thus further completing the division between the aorta and the pulmonary arte at the same time the two sets of 3 semilunar valves develop from the origin cushions (Fig 8)

8 Diagram to show the mode of formation of 2 sets of 3 semi lunar valves from 4 endocardial cushions.



(iii) The formation of the right atrioventricular orifice

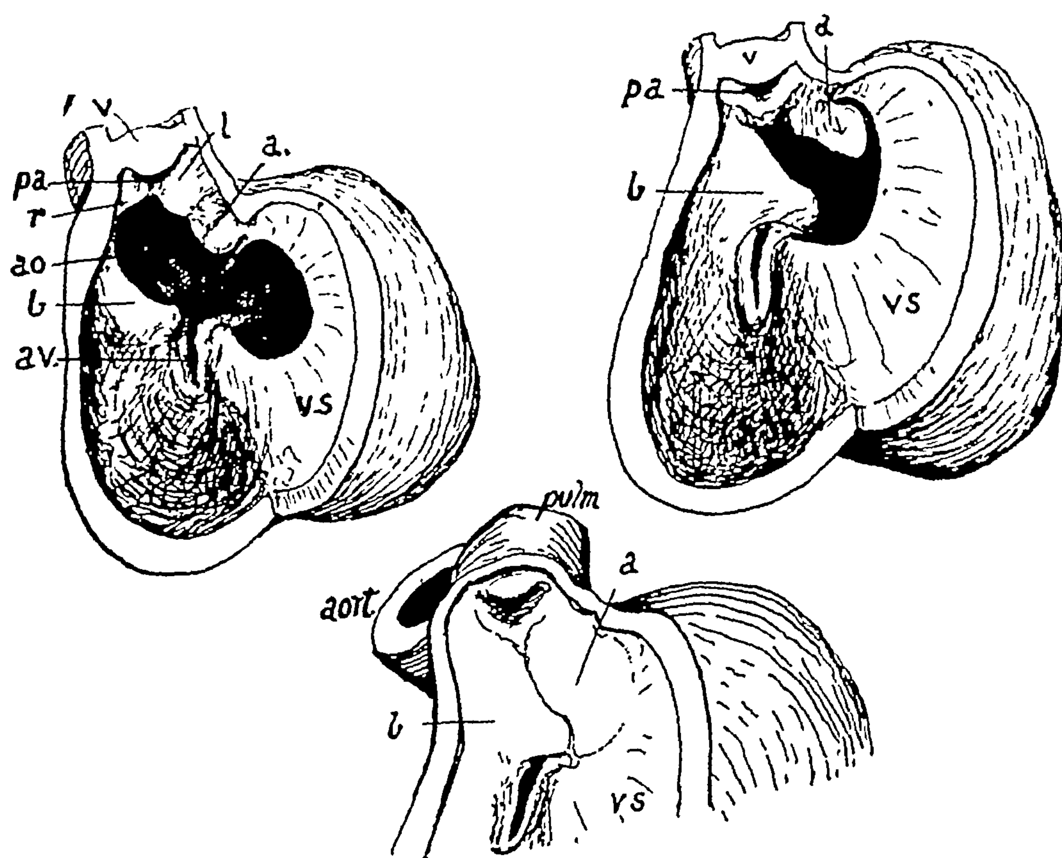
The angulation of the primitive heart tube brings the arterial and the venous components close together so that the formation of the two atrioventricular orifices is intimately associated with the closure of the ventricular septum and the development of the infundibulum. Two endocardial cushions (a ventral and a dorsal) form in the primary atrial canal, these cushions fuse and form the septum intermedium, thus separating the right from the left atrioventricular orifice. The ventricular septum reaches both the ventral and the dorsal endocardial cushions making a free falciform edge between them below the septum intermedium. It meets the dorsal endocardial cushion near its right extremity

(iv) The closure of the ventricular septum

The description has so far provided for two ventricles still partly connected above by a foramen which represents the lumen of the primitive heart. Above this foramen the truncus arteriosus has been divided to give the two great vessels with their semilunar valves, and behind it are the two atrioventricular orifices The closure of the ventricular foramen and the joining up of these somewhat diverse parts is accomplished chiefly by the *right and left bulbar ridges*, which form the proximal bulbar septum, and which develop in the walls of the bulbus below the distal bulbar septum, these two structures are of great importance to an understanding of the anatomy of the musculature of the normal and malformed right ventricle.

The two bulbar ridges (proximal bulbar septum) fuse above with the distal bulbar septum, which has already been mentioned as fusing above with the right and left arterial cushions, the separation of the pulmonary from the aortic orifice is thus continued further downwards.

The right bulbar ridge grows towards the left across the right atrioventricular orifice and just reaches the attachment of the ventricular septum to the dorsal endocardial cushion. The left ridge, in contact from its early stage with the upper part of the septum, begins extending along the free margin of this (Frazer, 1931) In this way the left ridge reaches the right ridge by growth along the edge of the ventricular foramen, and by fusion between the two ridges the aortic channel is roofed in and entirely separated from the pulmonary channel (Fig 9) The final closure of the foramen comes from a downgrowth of a small portion of the dorsal



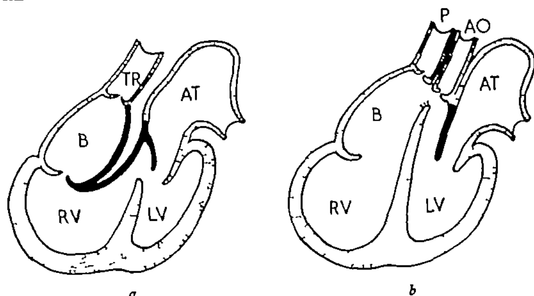
9 Diagram after Frazer to show mode of closure of ventricular septum by the bulbar ridges which develop from the proximal part of the bulbus (infundibulum)

pa Pulmonary artery
ao Aorta
a and *b* Bulbar ridges
vs Ventricular septum
av Right atrioventricular valve
l, *r* and *v* Left, right and ventral bulbar cushions which develop from the distal part of the bulbus (the arterial root) and form, with the dorsal cushion, the aortic and pulmonary valve cusps

endocardial cushion, this is the *pars membranacea septi*. This also intervenes between the right atrium and the left ventricle, i e. it is in part an atrioventricular septum.

(*v*) *The fusion of the bulbus cordis into the right ventricle*

Figure 10*a* shows diagrammatically the form of the embryonic human heart in the fourth week, Figure 10*b* depicts how by the third month the bulbus cordis has been incorporated into the first part of the truncus and into the wall of the right ventricle to form the infundibulum As can also be seen a small part of the bulbus is incorporated in the subvalvar part of the left ventricle and may be responsible for the maldevelopment known as congenital subaortic stenosis. For the purposes of this monograph this part of the bulbus which fuses with the left ventricle will henceforth be ignored

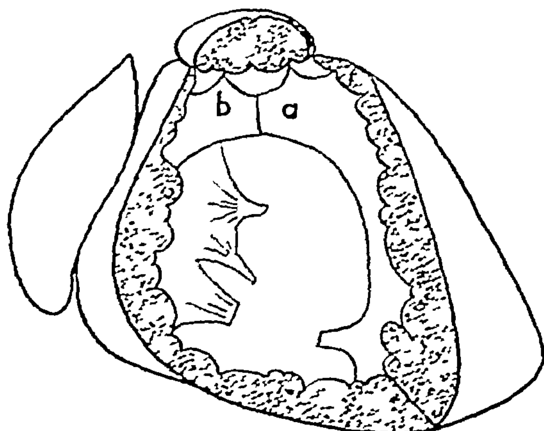


10 Diagram after Keith showing—on the left—that the bulbus is at first separated from the ventricle by the bulboventricular spur (in black) Later the spur regresses so that the proximal part of the bulbus is absorbed into the right ventricle to form part of the infundibulum—shown on the right.

Waterston (1918), in discussing the part played by the bulbus cordis in the formation of the right ventricle, states 'Perhaps the most striking change is the dilatation of the channel leading to the pulmonary artery Up to the 12 5 mm. stage the lumen was narrow and inconspicuous Now it is large and dilated and the orifice into the pulmonary artery is placed upon the dorsal wall, not at the termination. The ventral wall is lined with a reticulated formation of muscle continuous with that of the ventricle ' Thus the infundibulum of the right ventricle is formed from a portion of the bulbus cordis and is at first merely a narrow channel but becomes distended.

In the process of development of the ventricle the prominent bulboventricular spur to begin with causes the opening of the bulb to lead out of the right extremity of the ventricular cavity Later this spur becomes less and less prominent due to atrophy and disappearance of the dorsal and left wall of the bulbus until eventually the whole left and back wall of the bulbus have practically disappeared (Frazer 1931)

The chamber of the bulbus cordis, separate at an early stage, becomes overwhelmed by the musculature of the ventricles, it is not obliterated but is incorporated as an intrinsic part of the ventricular system. Keith (1924) states that 'the muscle cells of the developing heart have power of invasion of spreading into and occupying endocardial tissue masses and of grouping themselves into elabor ate formations In an earlier lecture (1904) he describes how the closure of the ventricular foramen is due to development of a great band of musculature which is characteristic of the mammalian right ventricle and of which no notice has been taken When the septal wall of the infundibulum of the right ventricle is examined there will be observed a raised, wide flat band of muscle fibres which



11 Diagram after Keith to show the formation of the crista supraventricularis by fusion of the right and left septal bands

end in the base of the aorta That band is well known in the reptilia in which it is attached above to the septum between the orifices of the aorta and pulmonary artery . . . It separates the arterial from the venous blood during systole of the reptilian ventricles ' This arrangement has already been described in the turtle's heart in Figure 5 and the reason for mentioning these muscular ridges within the single reptilian ventricle becomes clear when it is realised that they are significant in relation to the developing mammalian heart

Keith (1924) describes how the musculature of the developing right ventricle contains two large septal bands (the right and left, *a* and *b* in Fig 11) which invade and occupy the endocardial cushions of the right and left bulbar ridges The two bands unite to form the muscular structure known in the fully formed heart as the crista supraventricularis. An irregular linear raphe can be seen where these bands fuse, this line is only poorly shown in the normal heart but can sometimes be recognised in a malformed heart such as that shown in Figure 40 (p. 55) which is from an example of Fallot's tetralogy

The two muscular bands are continued from the crista supraventricularis along the right lateral and the septal wall of the right ventricle, in a manner to be detailed later, in such a way as to form a muscular constriction at the junction of the body and infundibulum of the right ventricle, a fact to which Peacock drew attention (1886). He pointed out also that it indicated the mode in which the right ventricle is developed, as illustrated in the heart of reptiles such as the turtle with its single ventricle imperfectly separated into three compartments by muscle ridges In systole this muscular constriction becomes more pronounced and indicates how, if some failure of expansion or imperfect inclusion of the bulbus cordis occurs, an obstruction can, and in fact does, occur at this level

CHAPTER III

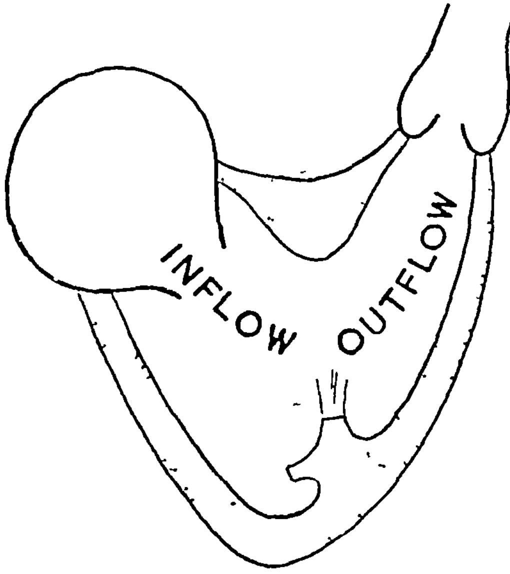
The anatomy of the normal right ventricle

BEFORE describing the details and varieties of pulmonary stenosis it is necessary to describe the structure of the normal right ventricle which is much more complicated than would ordinarily be supposed The ventricle consists essentially of two portions, an afferent inflow or atrial part into which the atrioventricular orifice leads, and an efferent or outflow part which leads into the pulmonary artery The first part is sometimes called the sinus and the second part is the infundibulum (Fig 12); they are normally separated by the muscular ring mentioned above which is formed by the crista supraventricularis and its continuations leading finally to the moderator band.

The formation of the right ventricle is also displayed in the postero-anterior and lateral angiocardiograms shown in Figures 13 and 14 The filling defect' caused by the downward projection of the wedge like crista supraventricularis is clearly seen in Figure 13

The key to understanding the right ventricle is the crista supraventricularis; this important muscle mass is depicted in Figure 15 which is a dissection from a sheep s heart. It is also seen in Figures 38 and 40 It forms a thick prominent ridge which projects down between the inflow and outflow portions of the ventricle. Its formation by fusion of the right and left septal bands was shown diagrammatically in Figure 11 and this arrangement is clearly indicated in Figure 40 (p 55)

To its right side the crista supraventricularis is continued on the anterior wall



12 Diagram of the normal right ventricle to show the inflow and outflow tracts

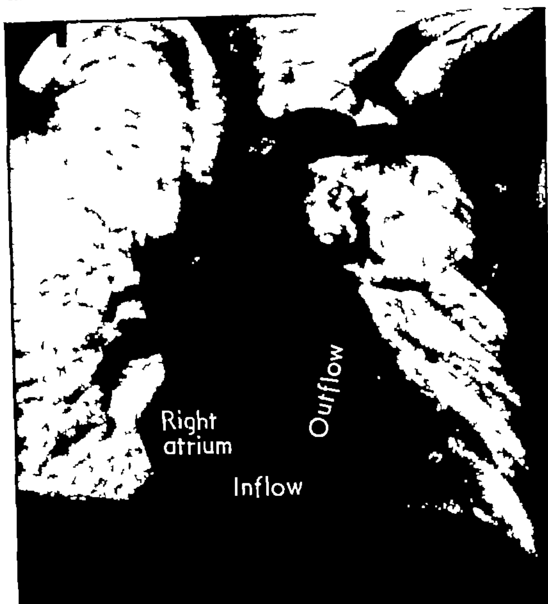
of the right ventricle towards the right border of the heart, its main branches run along the base of the ventricle on the lateral margin of the right atrioventricular orifice In systole it helps to draw the lateral against the septal wall of the ventricle and forms part of a safety-valve mechanism of the tricuspid orifice (Keith, 1904c) On the left the muscle sweeps as a wide band down the septal wall to end in the trabecular muscle of the apical region, the moderator band is part of this musculature and serves to bind it to the anterior papillary muscle The exact relations of the moderator band to the papillary muscle and their embryological significance are discussed by Mall (1912), who states that according to its development it should end in the site of origin of the anterior papillary muscle

The right limb of the atrioventricular bundle passes down the septum and along the moderator band and Keith (1924) states 'all of the right ventricle which lies above the main atrioventricular bundle and in front of its right branch is bulbar or infundibular in origin'

Behind the septal end of the crista supraventricularis and between it and the anterior cusp of the tricuspid valve is a small recess of variable depth which, although apparently insignificant, is of great developmental interest as it may, perhaps, represent the outflow channel of the right reptilian aorta

The infundibulum (Latin—funnel) stands out as a funnel-like portion of the right ventricle as can be seen when it is viewed from above, after removal of the atria (Fig 16) In this specimen the formation has been thrown into greater prominence by removing the superficial encircling fibres which enclose both ventricles This leads us to a consideration of the arrangement of the muscle fibres of the anterior wall of the infundibulum

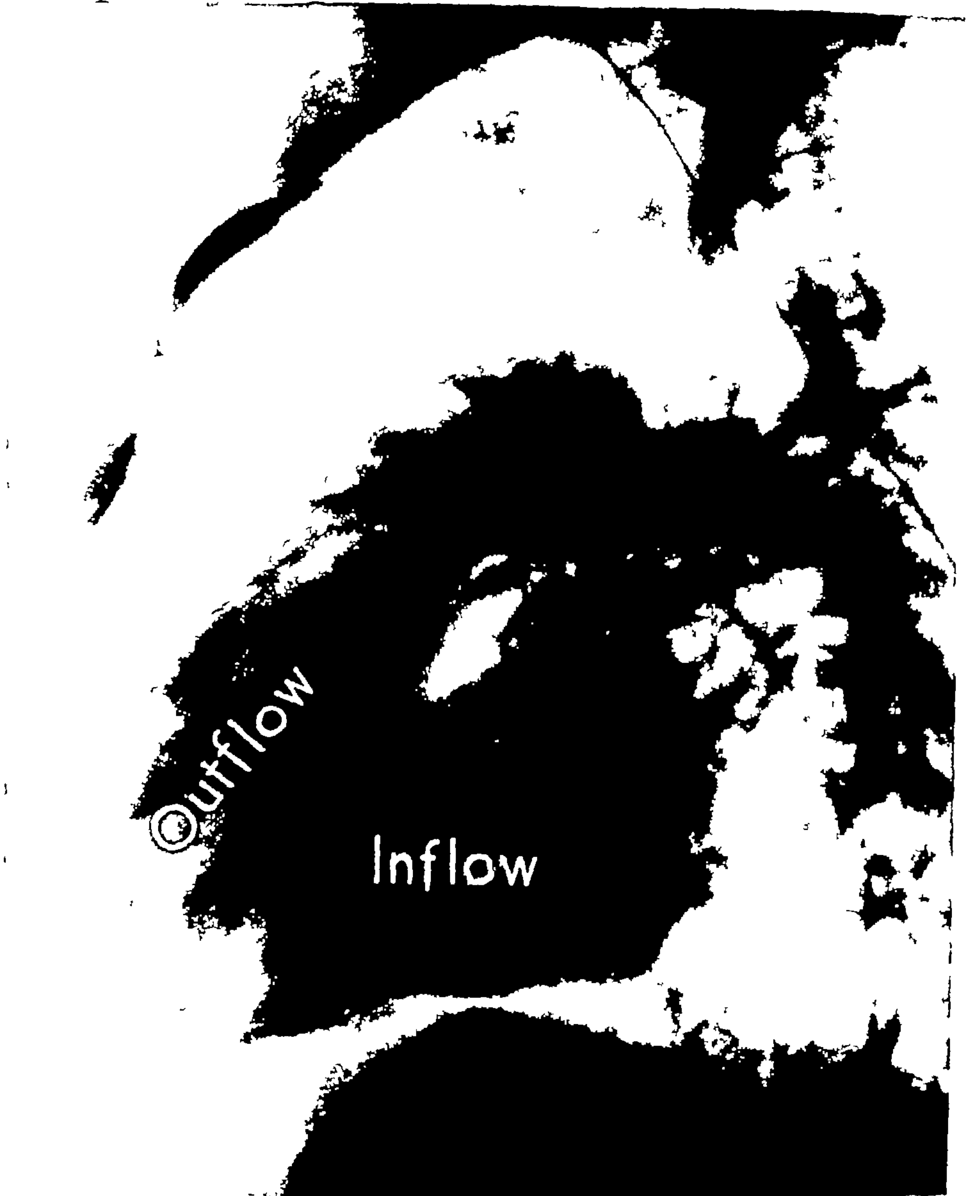
The superficial fibres consist of transverse loops which continue over both ventricles When these are peeled off a layer of transverse fibres is seen encircling the infundibulum alone (Fig 16) On the inner aspect of the infundibulum the fibres run longitudinally (Fig 17), this obeys the general rule pointed out by



13 Postero-anterior angiocardiogram to show the composition of the right ventricle

Ludwig (1849) that each piece of the ventricle which possesses the entire thickness of the wall has a system of fibres on its outer surface and a system at approximately right angles to it on its inner surface; and between them a series of layers in a regular succession of transition of direction from the one to the other (Quain, 1929).

This arrangement is of considerable practical significance in regard to the direction of surgical incisions into the anterior wall of the infundibulum If a longitudinal incision is made the more superficial, circular fibres are cut across transversely and tend to gape On the other hand the inner wall is cut in the direction of its fibres and when the wound is sutured there is no tendency to gape Scrutiny of the inner wall of the infundibulum after a longitudinal incision

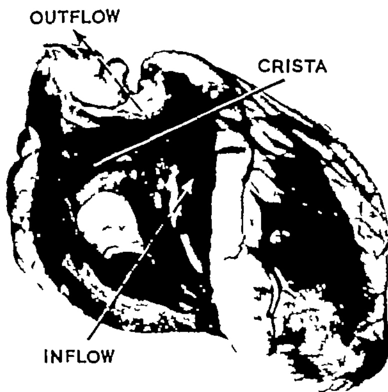


14 Lateral angiocardiogram to show the composition of the right ventricle.

has been sutured reveals that the line of closure is almost invisible, a fine, firm scar results and thrombus formation is minimal or absent If a transverse incision is made there is definite gaping along the inner surface of the suture line and inspection of the wound 7–10 days later in the heart of an experimental animal shows an obvious line of granulation tissue marking the irregular furrow There is no question, therefore, of the desirability of using a longitudinal incision in the anterior wall of the infundibulum

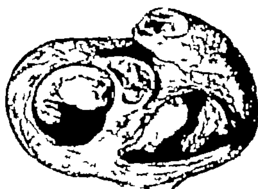
It is opportune here to mention briefly the question of nomenclature, about which there is often confusion Some mention has already been made of it in regard to the term bulbus cordis

The crista supraventricularis (B N A) according to Walmesley was first



15 View of sheep's heart to show the way the crista supraventricularis serves as a kind of watershed to separate the inflow from the outflow tract.

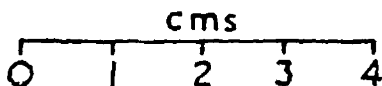




16 A sheep's heart from which the superficial thin investing layer of muscle fibres has been removed to demonstrate more clearly the structure of the infundibulum. The atria have been removed.



17 To show the longitudinal direction of the inner layer of muscle fibres of the infundibulum



accurately described by Wolff (1871) and although not definitely named by him, is usually called *éperon de Wolff* by French writers, Pettigrew names it the 'fleshy pons' and Sappey, Cruvelhier, and Sée the 'muscle compresseur de la valvule tricuspidale' (Quain, 1929) It should be noted that Keith does not use the term but describes the muscle mass as being formed by the two septal bands which he describes as 'a' and 'b', although Quain identifies Keith's band 'b' as being presumably the same as the crista supraventricularis

Quain also states that Wolff applied the term conus arteriosus to the arterial part of the right ventricle, a term which is still much used today although often loosely and vaguely in relation to a 'conus area', identified radiologically, which includes portions of the heart not certainly identifiable as this part of the right ventricle and which are not infrequently something entirely different, such as the pulmonary artery or the left auricle The term 'infundibulum', according to Quain, was first used by Sappey, Cruvelhier and Poirier, Sée names it the pulmonary canal Infundibulum is preferred in this book and will be used throughout in preference to conus arteriosus

Structure of the arterial root

Before leaving the anatomy of the right ventricle it is important to draw attention to the account in Quain (1929) of the structure of the 'arterial root', the

derivative of the distal part of the bulbus cordis, as it is relevant to some of the more distal malformations in this region Quain states that at each of the arterial openings there is a short tubular zone formed of fibrous tissue, the proximal borders of which, at the junctions with the ventricular muscle and with the typical arterial wall, are uneven.

This zone is a derivative of the bulbus cordis, as are also the semilunar valves which are formed on its inner surface Since the valves are derived from the proximal ends of the distal bulbar swellings, it follows that the part of the fibrous zone which lies between the attachments of the valves and the lower end of the typical arterial wall is derived from the distal part of the bulbus, the irregularity of its distal margin is due to the uneven advance of the arterial wall into the bulbus wall The part of the fibrous zone which lies proximal to the valves is the rudiment of the ventricular part of the bulbus, the muscle tissue of which has disappeared, the proximal part of it is included in the heart and covered with ventricular muscle, but the boundary between the two parts is irregular on account of the irregular distal excursion of the ventricular muscle.

'The boundary between the ventricle and the arteries, in the functional sense, is easily defined as the line of attachment of the semilunar valves It will be apparent, however, from what has been said above, that the morphological boundary by no means corresponds to the functional boundary· that the ventricle cannot be considered to extend upwards as the *spatia intervalvularia* nor the artery to extend downwards as the sinuses of Valsalva. Henle was the first to draw attention to this difficulty of defining the ventriculo-arterial boundary and introduced the term "arterial root" (the fibrous zone mentioned above) to replace the term "arterial ring" by which this boundary had been previously defined, and he described the unevenness of the margin of the ventricular muscle in a general way that it reaches highest in the spatia intervalvularia and that at certain places it reaches above the valve attachment. Luschka, however, was the first to describe the fibrous zones and their connections in detail

'The wall of the sinuses of Valsalva then, consists in part of the arterial wall and, at a varying distance below this, of the fibrous tissue of the "arterial root", in which there is little or no elastic tissue, and which is continuous with the fibrous tissue of the cusps of the valves at their insertion and with the fibrous tissue of the ventricular musculature At the transition of the artery into the "arterial root" the elastic tissue of the arterial wall becomes gradually less and ends in a pointed projection which lies close to the intimal coat

'The level of the line of transition from the artery to the arterial root is the same in all three sinuses of the pulmonary artery, the distance between the valve attachments and the arterial wall being the greatest at the middle of the sinuses, where it is about one third of the length above the cusp or margin and least at the sides The upward extension of the ventricular musculature is greatest in the left sinus of the pulmonary artery the greater part of the cusp at this sinus being attached to muscle tissue; in the posterior pulmonary sinus the musculature reaches just to the attached bands of the cusp, while in the right sinus it reaches

a little beyond it Langer has stated that in the child the attachment of the cusps is higher and is surrounded by ventricular muscle '

These rather long extracts have been quoted because they introduce and describe an anatomical and embryological matter of great relevance to the malformation of this region If it is a true hypothesis that the stenotic lesions arise from some imperfect development of the bulbus cordis, then it is just as important to know the distal limits of this structure and its level of junction with the truncus arteriosus as to know its proximal connection with the right ventricle since the distal part of the bulbus is absorbed into the truncus In this lies the explanation of pulmonary valve stenosis and of the forms of pulmonary atresia affecting the beginning of the pulmonary artery

The various malformations known collectively as pulmonary stenosis will be dealt with in the next chapter Before passing on to this it is as well to summarise the main argument and conclusions in connection with the embryology and surgical anatomy of the right ventricle

Summary

The right ventricle consists of two portions, a proximal or inflow portion—the sinus, a distal or outflow portion—the infundibulum Between these two parts is a muscular constriction formed, in the roof, by a large projection, the crista supraventricularis; important connections pass from this to the right wall and to the septal wall and so down to the apical musculature The infundibulum is developed from the bulbus cordis which, first of all a narrow passage, enlarges or expands and is then overwhelmed by a growth of the ventricular muscle into it. The stage of formation of the bulbus cordis and its fusion into the right ventricle and the truncus arteriosus is seen for a short time in the human embryo A well-formed bulbus cordis is a normal constituent of the heart of the gill-breathing fish. Distally the bulbus tissue forms the pulmonary valves and first part of the pulmonary artery, the so-called arterial root The development of the infundibulum and the inclusion of the bulbus cordis is intimately concerned with the closure of the ventricular septum

It remains to be shown how the majority of examples of malformation associated with pulmonary stenosis are due to some abnormality of the expansion, development or fusion of the bulbus cordis

CHAPTER IV

The causal relationship of the pulmonary stenosis and the septal defect

Before describing the various types of pulmonary stenosis it is as well to discuss the question of the relative significance to each other of the septal defect, the dextroposed aorta and the pulmonary stenosis, in other words, which is likely to be the primary lesion?

Peacock (1866) has discussed this matter in an interesting and informative manner Fallot's clinical description of the malformation associated with his name was written some 22 years later (1888), and it will be seen from the following description that he was anticipated by Peacock who writes, In one of the most interesting forms of anomaly the deviation of the septum is to the left, so that the right ventricle is of large size, and the aorta arises wholly or in part from that cavity· and this condition is most generally associated with obstruction to the passage of the blood from the right ventricle. He then discusses which is the likely primary lesion and his remarks are so pertinent that they are best quoted in full a step which has the additional merit of making some extra acknowledgement to the value of his contribution. After pointing out that Meckel favoured the septal defect and displacement as the primary lesion whereas William Hunter (1812) thought the pulmonary obstruction to be the exciting cause, he agrees with Hunter and continues, Hunter pointed out that if during early foetal life when the interventricular septum was incomplete some obstruction arose by which the right ventricle became incapable of freely transmitting the blood which

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18 Thomas Peacock

it contained through the pulmonary artery into the ductus arteriosus and descending aorta, the current must necessarily pass through the aperture in the septum into the left ventricle and thus the final separation of the two cavities would be prevented To these views it is scarcely possible to object and it seems equally to result from them that the septum in its further growth might be made to deviate to the left, so that the aorta would communicate with the right ventricle It also follows that from the connection between the aorta and the right ventricle, the constant flow of blood from the cavity into the vessel must tend to draw the aortic orifice still further to the right, so as to produce widening of the aperture and of the ascending aorta which is so peculiar a feature in all those cases in which that vessel had a double origin from the two ventricles

Meckel suggests that incompleteness and displacement of the septum are primitive defects, and that in consequence of the blood finding a ready outlet from the right ventricle through the aorta the pulmonary artery becomes more or less abortive, and the orifice is contracted down during the progress of development. This view however throws no light on the mode of formation of those very much more numerous cases in which, with the diminution in the capacity in the orifice and vessel the valves of the pulmonary artery are seemingly diseased. It leaves also the nature of the original defect unexplained, and substitutes for what appears in many cases an obvious and sufficient cause, an entirely theoretical idea.

CHAPTER V

The varieties of congenital pulmonary stenosis

CLASSIFICATIONS are apt to recoil on the head of their author, especially when knowledge and experience are incomplete However, it is necessary to attempt some logical arrangement, if only to make description and discussion easier The classification and comments that follow are provisional and dependent on present experience

The varieties of pulmonary stenosis to be discussed are

(*i*) *Pulmonary valvar stenosis with normal aortic root*

a. with closed atrial septum
b with interatrial communication

(*ii*) *Infundibular stenosis with normal aortic root*

(*iii*) *Pulmonary atresia*

a arterial atresia
b valvar atresia

(*iv*) *Tricuspid atresia*

(*v*) *Fallot's tetralogy*

a valvar stenosis

b infundibular stenosis
c. combined valvar and infundibular stenosis
d septal defect.

(*vi*) *Aorta and pulmonary artery both arising from right ventricle with pulmonary stenosis, and septal defect*

(*vii*) *Transposition of the great vessels*, with pulmonary stenosis

(*viii*) *Common ventricle with pulmonary stenosis*, fusion of inflow portion of right ventricle with left ventricle, semi vestigial infundibulum.

This monograph is concerned primarily with a description of the anatomy of the various types of pulmonary stenosis, but such a description is enhanced by reference to radiological and clinical features and to operation findings The need for an anatomical description rests on clinical application and it is therefore timely to say something of the recognition of the various types of stenosis

This does not imply a full clinical description and differential diagnosis but merely some general remarks on the problem.

In most types a firm diagnosis can be made before operation by consideration of the clinical features, physical signs, electrocardiography and the full range of radiography, including angiocardiography; cardiac catheterisation increases yet further the extent of preoperative diagnosis But even with these aids and with considerable practical experience it may still be difficult or impossible to make a firm or complete diagnosis before operation

Moreover, it may not be desirable or worthwhile to apply all the available tests in order merely to obtain a complete anatomical diagnosis Thus, in most cases of Fallot's tetralogy, if the basic diagnosis is acceptable, it is but rarely necessary to use cardiac catheterisation and angiocardiography to try and elucidate the exact site and type of the stenosis Once the need for operation has been accepted the opportunity for making a precise anatomical diagnosis will present when the heart is exposed at thoracotomy The direct information obtained will be much more exact than that offered by indirect preoperative methods The actual plan of operation is not affected by the type of stenosis even if this is known beforehand

In cases of presumed pulmonary atresia it is only at operation that the exact observations can be made that decide whether or not a palliative operation is possible.

In other words, exposure and examination of the living heart is the best final method of obtaining a diagnosis In most cases it is superior to post-mortem study for, although the total exposure at autopsy may permit a more complete examination of some parts of the heart, post-mortem examination, in general, suffers from the grave disadvantage that it is done on a dead structure which is often in rigor mortis, may be hardened by formalin, and frequently deformed or distorted in the process of fixation. This is the main reason why so much of

what has been written and taught about pulmonary stenosis is inaccurate and misleading

When the heart is exposed at operation, with the pericardium opened widely, much can be learned by inspection, palpation (both external and internal), by insertion of instruments and by electromanometric measurements made in the various chambers and vessels both by needle puncture and by catheter As will be shown in the following descriptions it is possible to make a full diagnosis of the site, nature and severity of the pulmonary stenosis except in rare cases

We have, in fact, another example of the great advantages of a study of the pathology of the living

CHAPTER VI

Pulmonary valvar stenosis with normal aortic root

(syn. pure pulmonary stenosis, 'Trilogy of Fallot')

The term 'Trilogy of Fallot' is popular in the French literature but is imperfect; not entirely satisfactory is 'pure pulmonary stenosis' which, though a convenient term to indicate absence of ventricular septal defect and an over riding aorta, fails in those cases in which there is a communication between the atria. Allanby and Campbell (1949) have given a valuable report on the condition, its operative treatment has been dealt with by Campbell and Brock (1955) and by Milstein and Brock (1955). A few years ago it was thought to be a rarity but now that the diagnostic features have been recognised it is accepted as fairly common. Thus Campbell (1953) in an analysis of 1 100 cases of congenital heart disease at his clinic found that its frequency was about equal to that of persistent ductus arteriosus

The valvar obstruction

The basic feature of the stenosis is fusion of the cusps of the pulmonary valve to form an obstructing membrane which under the influence of the right ventricular pressure, projects as a dome-shaped or conical structure into the pulmonary artery the exact shape is doubtless influenced by the thickness or thinness,

19 Photograph of severe pulmonary valve stenosis The very dilated pulmonary trunk has been opened to display the obstructing valve dome



a *b* *c*

20 Photographs of three types of pulmonary valve stenosis (For details see text)

rigidity or suppleness of the fused valve cusps Although it is natural for the obstructing membrane to become thicker and more rigid as the years advance, it may be found to be short, thick and rigid even in children. In older patients small warty vegetations are often found around the small central orifice on the arterial aspect, occasionally the apex of the valve cone is lightly calcified. The lumen, in extreme cases, may be no more than 2–3 mm in diameter (Fig 19), and it is remarkable that life can continue when it is realised that the whole of the circulating blood has to pass through such a tiny aperture. The lumen may, in milder cases, be as much as 0·5–1 cm. in diameter It is generally possible to recognise the three cusps and their fused commissures (Fig 20*a* and *b*), occasionally the distal surface is smoothed out with no trace of the cusps (Fig 20*c*)

It is still often stated that the fusion of the cusps is due to 'foetal endocarditis, a conveniently vague term which could be accepted if it definitely included non bacterial processes

The chief difficulty in understanding valvar stenosis developmentally has been the apparent fusion of three well-formed cusps in a heart in which the development of the ventricular septum has been completed. Clearly it has occurred much later than the malformation in Fallot's tetralogy which must date from before the heart is fully formed

Keith states 'under the microscope the valves are seen to be composed of dense laminated fibrous tissue at their margins, but the body of the valves shows a reticulated tissue distinctly embryonic in character'

It is not unreasonable that such a valvar obstruction can occur as a predominant feature in a maldevelopment of the distal part of the bulbus cordis, from which it should be remembered, the valves develop The specimen in Figure 35 (p 49) is of great interest in this connection for, although from an example of pulmonary atresia, it shows the valve cusps are well formed but fused and there is complete obliteration of the channel immediately below them, the ventricular septum is fully formed.

The closure of the ventricular septum depends upon the development of the more proximal connections of the bulbus cordis and it is not difficult to accept that the valves should be malformed and yet the rest of the structures developed from the bulbus cordis show only slight abnormality

The right ventricle

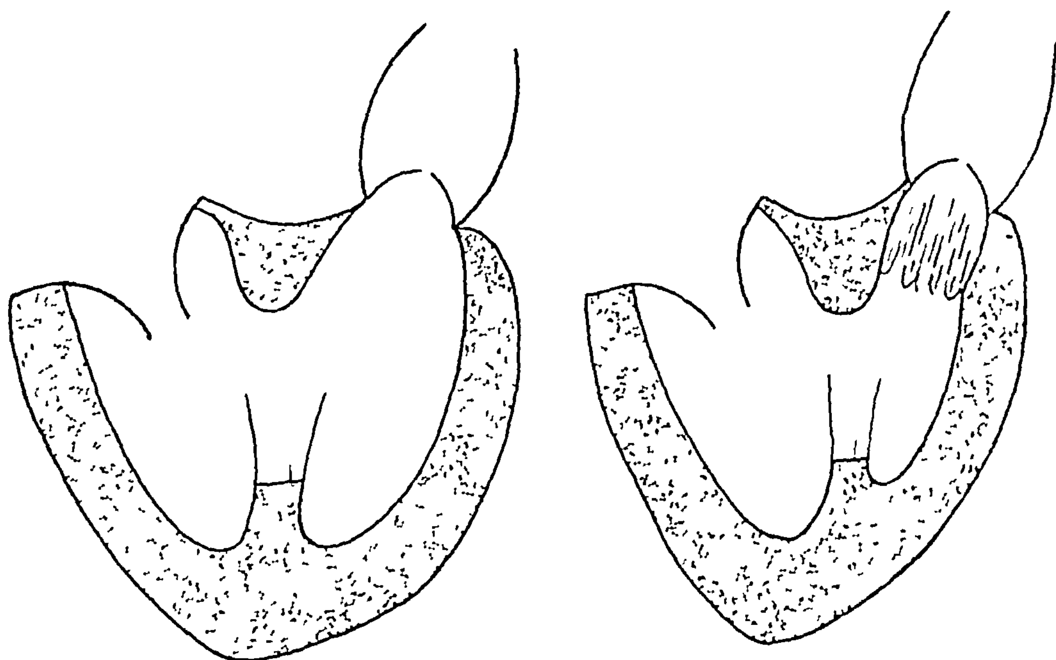
Abnormality of the other structures developed from the bulbus cordis although slight, are however important for examination of the right ventricle in these cases of 'pure pulmonary valvar stenosis reveals significant features which clearly indicate the condition is an earlier developmental one rather than a later acquired state.

In addition to the valvar fusion, the infundibulum is smaller and shorter than it should be, usually the infundibular lumen, even though smaller, is adequate except in the circumstances to be described later Moreover the infundibulum

exhibits the same 'icing' lining of thickened opaque endocardium seen in frank infundibular maldevelopment as in Fallot's tetralogy. Today it is customary to dignify this state by such terms as 'endocardial fibroelastosis', etc.

Again, although the lumen of the pulmonary artery above the valve is always dilated, and often grossly so, the actual orifice of the artery at the level of the attachment of the valves may be small, much larger than the pin-hole opening in the valve and not constituting an obstruction but nevertheless smaller than is usual in a patient of like age After successful valvotomy this secondary stenosis at the pulmonary ring may assume a greater importance

In Fallot's tetralogy, when a high-grade valvar stenosis is present, the infundibulum is often poorly developed and recessive and so cannot partake fully,



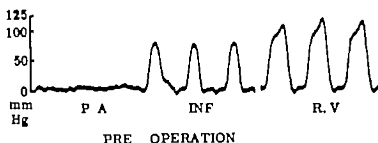
21 Diagram to show how the outflow tract is abnormal in pulmonary valve stenosis

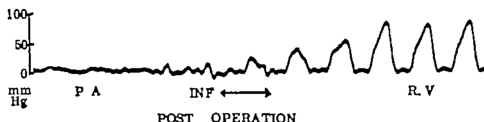
especially in the presence of a ventricular septal defect, in the hypertrophy and dilatation that normally occurs proximal to such an obstruction. In 'pure' pulmonary valvar stenosis the infundibulum is only slightly affected and its muscle is well developed so that it can hypertrophy with the rest of the ventricle to compensate for the obstruction Commonly, therefore, any subdevelopment of the infundibular muscle is overshadowed and the only stigma that may remain is the endocardial abnormality and a change in form (Fig 21)

The hypodevelopment of the infundibulum may, however, become important when it is complicated by excessive muscular hypertrophy, for in this way a secondary or associated infundibular stenosis may be added to the valvar stenosis. This is a phenomenon I have been observing for some time and is exemplified by the electromanometric tracing shown in Figure 22 The upper tracing was taken at operation and immediately before pulmonary valvotomy, it shows that

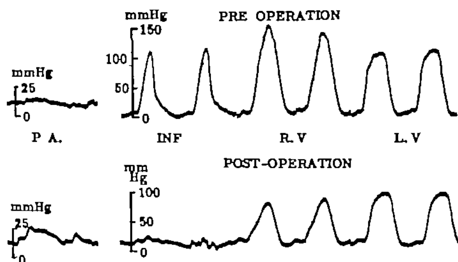
the pressure in the pulmonary artery is very low and has little wave form. The pressure in the infundibulum rises to about 80 mm and in the body of the ventricle proper the pressure is about 115 mm.

A satisfactory pulmonary valvotomy was done and the lower tracing was then





22. To show the effect of pulmonary valvotomy on the production or aggravation of an infundibular stenosis



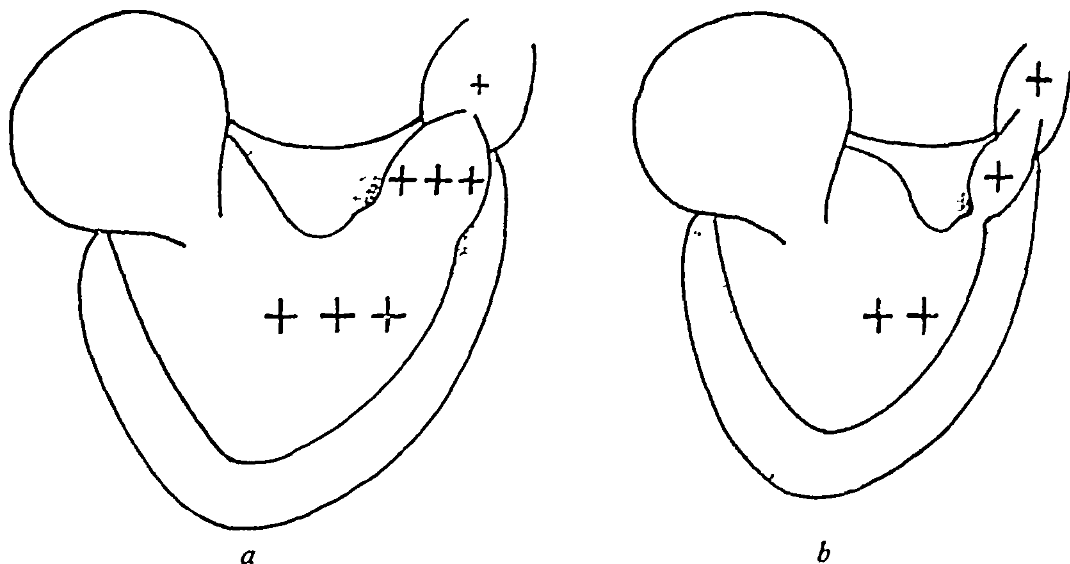
23 Another example of change in position of the gradient from valve to infundibular level after pulmonary valvotomy

obtained. It will be seen that the effect of the valvotomy has been to shift the major pressure change from the valve level back to the infundibulum, the right ventricular pressure is still about 75 mm.

Figure 23 shows the same phenomenon in another case. In the top tracing the presence of a dominant valve stenosis is clearly seen, in the lower tracing,

taken after valvotomy, there is now a single infundibular stenosis. In both these cases a finger was introduced into the ventricle and the presence of an infundibular stenosis was confirmed, the satisfactory relief of the valve stenosis was also noted The infundibular stenosis was then resected

The explanation is shown in Figure 24 In '*a*' is seen the state of affairs before valvotomy Some degree of infundibular narrowing is present but the obstructing valve barrier causes such a marked rise in pressure behind it that the walls of the infundibulum are held apart In '*b*' valvotomy has been done successfully and the obstructing valve membrane no longer exists, the infundibulum is, as it were, decompressed, its walls are no longer held apart but can come together and the infundibular stenosis appears Note that though the high pressure in the body of the right ventricle is lowered it is still above normal The previous high pressure

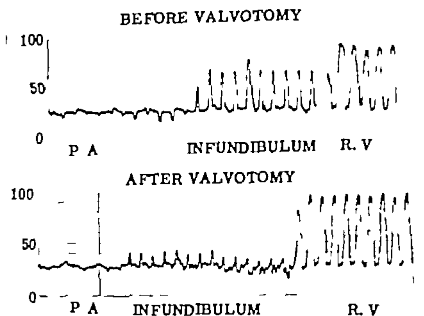


24 Diagram to explain the mechanism of change in level of gradient

in the infundibulum is replaced by a low pressure In other words valvotomy has resulted in a secondary infundibular stenosis appearing

When this phenomenon had been recognised I made a search through the tracings taken at operation in certain other earlier cases of pulmonary valvotomy and at once other examples were found Figure 25 is from one of these The upper tracing shows a severe pulmonary valve stenosis, a moderate infundibular stenosis The lower tracing shows a typical infundibular stenosis with no relief of pressure in the right ventricle. Inevitably this patient had a poor result

Several things can be learned from these observations Most, although not all, were seen in late, neglected cases, usually in patients in the twenties or thirties The muscle of the right ventricle is often enormously thick and there seems little doubt that this causes, or at least aggravates, the condition. In other words the heart is 'muscle-bound' The wall of the ventricle is so thick that it begins to fill up the outflow tract The truly enormous size is emphasised by the crista supraventricularis which can be felt by the exploring finger to form a thick wide ridge



25 A failed case of pulmonary valvotomy in which reference back to the operative tracing shows the aggravation of *infundibular* stenosis after valvotomy

of muscle perhaps 3 cm across so big that it is difficult to punch it with the jaws of the punches usually available

It may be that after the partial relief of the obstruction by valvotomy the gross hypertrophy of the muscle lessens and the outflow tract opens slightly more; the obstruction is thus further relieved and more muscular regression occurs so that progressive secondary improvement can happen over many months A benign circle is set up

In many, however, the condition seems very resistant and it appears to be almost an end result condition The possibility of its development is a powerful argument in favour of early operation in all cases in which the stenosis is at all significant. Any policy of delay in younger patients must take into account the later development of this secondary muscular obstruction in a heart that has been struggling against a stenosis for many years

There can be little doubt that this secondary infundibular stenosis following pulmonary valvotomy is responsible for many of the cases of failed valvotomy, or of the poor results said to follow the operation. Figure 25 is an example of this

Kirklin *et al* (1953) have also drawn attention, in an excellent article, to this association between valvar and infundibular stenosis in certain cases with intact ventricular septum. In addition to showing electromanometric tracings illustrating the aggravation or development of infundibular stenosis after valvotomy, they show photographs of three specimens in which the grossly hypertrophied muscle of the outflow tract is clearly seen forming an infundibular obstruction. They point out how this may be responsible for maintenance of right ventricular obstruction even after a completely adequate valvotomy They also quote a case in which, although a considerable degree of infundibular stenosis persisted after

valvotomy, cardiac catheterisation six months later showed a considerable fall in right ventricular pressure had taken place This indeed supports the suggestion that valvotomy may allow diminution of subvalvar muscular hypertrophy and thus progressive relief of the obstructed right ventricle.

Blount, Swan *et al.* (1954) have put forward a strong plea for open trans-arterial pulmonary valvotomy instead of the 'blind' transventricular operation They point out that their own experience and that of some surgeons in the literature is that it is not often possible to obtain complete or even adequate lowering of the right ventricular pressure or conversion of the gradient across the pulmonary valve by transventricular valvotomy They assume that this is inevitably due to limitations of the efficiency of the 'blind' procedure and that the valve stenosis is incompletely relieved In certain cases there is no doubt that even though valvotomy has been performed totally and completely, the development of the secondary muscular infundibular stenosis described above may prevent satisfactory relief of the obstruction and fall of the right ventricular pressure. An open valvotomy may give as perfect a valvotomy as it is possible to achieve, but it cannot correct or avoid this secondary infundibular muscle stenosis This may be corrected by open resection of the infundibular musculature, but it may not be good physiological surgery to do this

The great hypertrophy of the musculature of the right ventricle forms one of the most prominent features in the external appearance of the heart It is usually paler than normal and only few superficial branches of the coronary vessels are to be seen, in contrast to cases of Fallot's tetralogy in which the muscle is usually darker and more vascular, the coronary vessels being sometimes large and tortuous and occupying much of the ventricular surface

In most cases the right ventricle enlarges over to the left, pushing the left ventricle backwards so that this chamber is often quite concealed in a front view. This means that with the usual left anterior thoracotomy incision the right ventricle is the most prominent structure in the operation field In rare cases (2–3 per 100) there is anticlockwise rotation and the right ventricle may be concealed beneath the sternum

In addition to enlarging to the left, the right ventricle enlarges upwards and forwards so that it comes to resemble a pouter pigeon's chest This has an important effect in that it carries the origin of the pulmonary artery with it so that the pulmonary trunk comes to lie horizontal or even, in extreme cases, inclines downwards and backwards, this may also cause severe distortion so that the axis of the main pulmonary artery no longer lies in the axis of the infundibulum; the Z-shaped junction favours secondary obstruction

The pulmonary artery

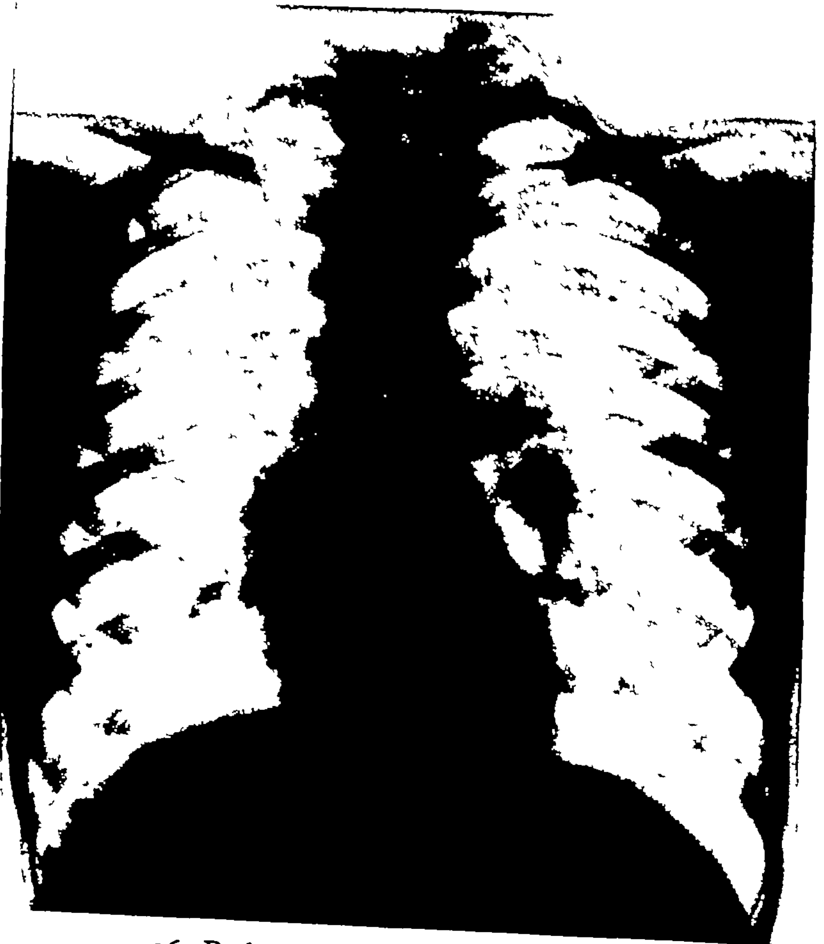
The post-stenotic dilatation of the pulmonary artery is a prominent and important diagnostic feature, it is an ovoid enlargement beginning at the level of the pulmonary ring and seeming to affect the whole trunk Actually not all the

circumference is involved as is seen when it is dissected, for the level of bifurcation is found to be quite low down, indicating that the dilatation is chiefly at the expense of the anterior and right and left lateral surfaces There is no trace of the sinuses of Valsalva which are smoothed out by the dilatation. In some cases the dilatation extends distally into the left branch (Fig 26).

The mode of development of the post-stenotic dilatation is still not completely understood for certain, although the observations and conclusions made by Holman (1954) are the most acceptable. He points out the well known physical fact that when a mass of fluid is ejected through a narrow constriction at high velocity it strikes against the more slowly moving fluid distal to the stenosis and is slowed down, its kinetic energy is converted into lateral pressure and its stream is deflected laterally or is even completely reversed so that eddies of alternating high and low pressure are set up, these cause repeated impacts over a long time and the elastic wall undergoes structural fatigue and distension. The lateral pressure effect is further enhanced by the hydraulic principle that a widening stream causes decreasing velocity but increases lateral pressure Holman has demonstrated convincingly the *onset of structural fatigue and sudden appearance* of aneurysmal dilatation by simple experiments on lengths of rubber tubing

The post-stenotic dilatation of the pulmonary artery provides an important diagnostic radiological feature when occurring in association with lung fields with diminished vascularity (Fig. 27). The heart is often rather globular except when it becomes grossly enlarged, it then becomes more angular (Fig 28*a*) In these extreme cases the dilated pulmonary artery is pushed so far upwards as to be quite concealed in the heart shadow, but is revealed in the angiocardiogram as a circular opacity, for the trunk is viewed end on and is no longer seen in its long axis (Fig 28*b*). The opacification of the artery may persist throughout the examination owing to the slow circulation in the vessel Figure 26 shows the usual ovoid dilatation of the main pulmonary trunk and also to an unusual degree the manner in which the post-stenotic dilatation may extend into the left pulmonary artery

When the pulmonary artery is examined at operation there is usually no doubt about the diagnosis The form of dilatation of the vessel is characteristic, the wall is felt to be under low tension and may be so thin that the dark blood is easily seen through it. A finger placed lightly on its proximal part notes the low tension and also detects the characteristic thin, jetlike thrill emerging from the valve opening With slight finger pressure at the base of the artery the tense cone of the valve can be felt in systole and if the finger is passed over it the origin of the emergent jet can be clearly felt Finally if the base of the artery is lightly held between the finger and thumb the whole conelike or domelike structure of the stenosed valve can be felt. It is sometimes possible to feel irregular fibrous or even calcified nodules around the small valve opening These last two manipulations must be used with great care and for only a very short time, otherwise a rapid fall in blood pressure will occur Although the diameter of the pulmonary artery is much larger than normal and is greatest at about its mid-length, the





26 Radiograph and angiocardiogram to show gross post-stenotic dilatation of the left pulmonary artery





27 To show post-stenotic dilatation of pulmonary trunk.

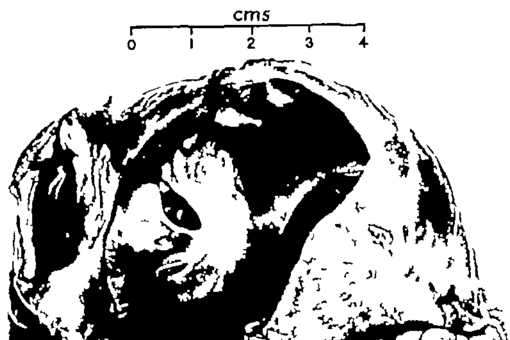


a



b

28 Huge heart in which the shadow of the dilated pulmonary trunk is concealed in the plain film but revealed in the angiocardiogram



29 Persistent foramen ovale seen in association with pulmonary valve stenosis

artery is often smaller than normal at its origin and indeed may be very much smaller As already stated, after a successful valvotomy this may even constitute a secondary stenosis at the pulmonary ring

The large hypertrophied right ventricle is matched by the large, dilated dark walled right atrium and its appendage, in contrast to it, in those cases in which the atrial septum is intact, the left atrial appendage is seen to be a small, lax pink structure

When there is a communication across the septum the left atrial appendage resembles the right appendage in size, colour and tension.

This communication is commonly small, often surprisingly so, and is usually a foramen ovale which has become incompetent as a result of the mounting pressure behind the obstructed right ventricle (Fig 29). Much more rarely there is a septal defect away from the fossa ovalis

CHAPTER VII

Infundibular stenosis with normal aortic root

(syn 'pure' infundibular stenosis, subdivision of the right ventricle)

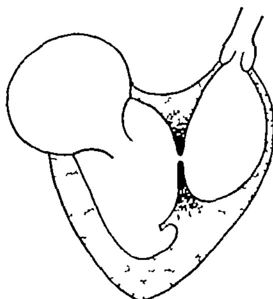
BOTH Peacock and Keith described examples of pure infundibular stenosis, both the atrial and ventricular septa being closed.

Peacock, after describing cases in which the development of the constriction or 'supernumerary septum' occurred before the division of the ventricle was completed, or at an early period of foetal life, then differentiates a type in which the abnormal partition is the only defect He describes a case 'In the cavity of the right ventricle there was a septum, dividing the sinus from the infundibular portion, this septum was perforated by an oval aperture 47 25 mm in circumference, by which the two divisions of the cavity communicated '

According to Keith and to Peacock a subdivided ventricle may be the only cardiac abnormality, there is no ventricular septal defect and consequently no dextroposition of the aorta It is simply that the normal zone of muscular narrowing which lies between the inflow and the outflow portions of the right ventricle is so over-developed that there is a muscular partition or septum (virtually a much overgrown crista supraventricularis) containing one, two or three openings (Fig 30) The edges of the opening are fibrous, if the lumen is reasonably large (e g 1–1 5 cm) the condition is compatible with long life and it may even be an incidental finding at post-mortem The smaller the stoma the worse the prognosis

Keith (1909a) states that the condition is 'comparatively rare' and found 19 hearts of this nature in the series of 270 malformed hearts which he examined It

30 Diagram of pure infundibular stenosis.



appears, though, that he included a number of cases which should really be classed as examples of Fallot's tetralogy with low infundibular stenosis and a large chamber For instance, amongst this 19 were nine with a patent ventricular septum and in four there was also a pulmonary valvar stenosis, in three the ductus arteriosus was persistent and in two the foramen ovale was still open. These observations indicate that not all were examples of a simple subdivision of the right ventricle.

Maude Abbott (1936) describes two cases of pure infundibular stenosis from amongst her 1,000 total. She writes, 'These cases are remarkable in that the seat of the stenosis was not at the pulmonary valve but some distance below this at the "lower bulbar orifice" forming a capacious separate chamber with a small orifice in its place.

Pure infundibular stenosis is not nearly so rare as is thought. Greater experience in cases of pulmonary stenosis with closed ventricular septum has led to the recognition and differentiation of a number of examples Thus I have myself operated upon 9 cases during the last 5 years, since 1948 I have operated on 111 cases of pulmonary valvar stenosis with closed ventricular septum

The diagnosis should be thought of when, in a case of pulmonary stenosis with closed septa, no post-stenotic dilatation of the pulmonary artery can be seen although the heart is not too big to conceal it; a bulge may be noted at the site of the infundibulum due to prominence of the chamber

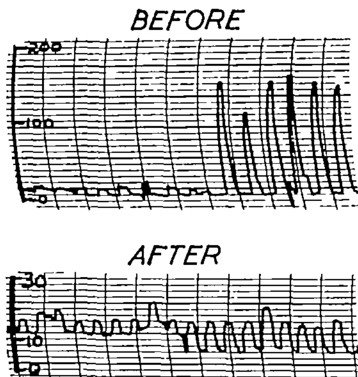
The angiocardiographic appearance is characteristic as can be seen from Figure 31, which shows the stenosis and the post-stenotic chamber clearly The prominence of the post stenotic chamber which enables it often to be recognised as a bulge even on a plain radiograph and which is a direct diagnostic feature at operation, is related to the thinness of the muscle in the outer wall. This may thin out so rapidly from just above the stenosis that the greater part of the outer wall of the chamber may be composed of little more than thin fibrous tissue



31. Angiocardiograms of pure infundibular stenosis

through which the blue blood is clearly seen. This poverty of muscle of course reflects the maldevelopment of the infundibulum as a whole, of which the actual stenosis is but one part.

Cardiac catheterisation also enables a firm diagnosis to be made from the characteristic pressure tracing obtained as the catheter is withdrawn from the pulmonary artery and through the infundibular chamber on its way to pass through the stenosis into the high pressure part of the right ventricle (Fig 32)



32 Electromanometric tracing before and after open resection of pure infundibular stenosis

The typical feature is the drop in diastolic pressure within the infundibular chamber as opposed to the pressure within the pulmonary artery

An important anatomical feature is that the papillary muscles and tendons of the septal cusp of the tricuspid valve arise from the proximal aspect of the mus cular ridge of the stenosis This close relationship makes it quite unsafe to do a closed blind punch resection of the infundibular stenosis, for organic tricuspid incompetence may be caused this I have caused in one case Since then I have performed open resection of the stenosis under direct vision when it has been easy to demonstrate the precision that is needed to avoid damaging the tricuspid valve mechanism

This close relation of the tricuspid valve to the stenosis means that when a catheter withdrawal pressure tracing is being made the catheter may pass directly from the low pressure of the infundibular chamber into the right atrium, only if

the catheter is then advanced again may the body of the right ventricle be entered and a high-pressure chamber demonstrated

Figure 33 shows the total resection of infundibular stenosis that has been possible under direct vision surgery. In Figure 32 can be seen pressure tracings made before and after operation, the obliteration of the pressure gradient shows that the obstruction has been completely relieved



33 Tissue removed at open resection of infundibular stenosis

CHAPTER VIII

Pulmonary atresia, tricuspid atresia

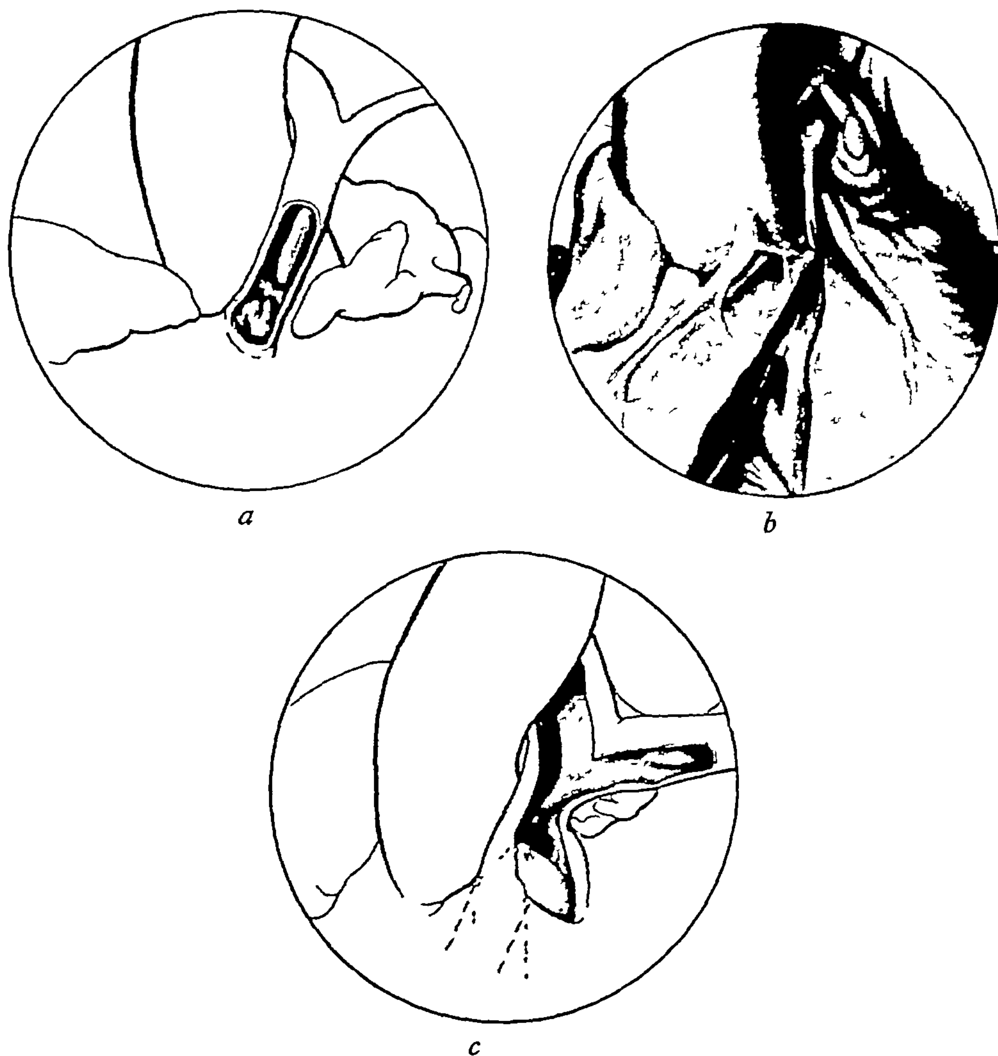
Pulmonary atresia

An excellent survey of this condition has been given by Allanby, Brinton, Campbell and Gardner (1950)

Arterial atresia

The pulmonary artery may be obliterated for a varying distance either just above the valves (Fig 34*b*) or throughout its length There may be no trace of the pulmonary stem or a fine fibrous thread may lead up to the horizontally running vessel formed by the right and left pulmonary arteries In these cases the circulation of blood within the pulmonary arteries is almost stagnant and is wholly dependent upon collateral circulation from the bronchial artery system, unless the ductus has persisted Sometimes the right or the left or even both pulmonary arteries may be obliterated

In cases of pulmonary atresia the surgeon must often explore the local anatomical arrangement in the hope of finding a pulmonary vessel large enough to use for a useful anastomosis to a systemic artery



34 Three examples of small pulmonary trunk and atresic valve (*After Allanby, Brinton, Campbell and Gardner*)

(*a*) The atresic valve consists of a mass of fibrous tissue in which no cusps can be made out

(*b*) The small size of the infundibulum and valve ring can be seen with the tiny pulmonary trunk, which widens just below its bifurcation

(*c*) Atresic valve seen from above, note the orifice is eccentric

Valvar atresia

This is a fairly common and important form of pulmonary atresia that is well described by Allanby *et al* (1950) The atresic valve can be recognised as consisting of two very imperfectly formed cusps fused together, often the atresic valve is eccentric, the cusps being fused obliquely to the arterial wall (Fig 34*a*)

The fibrous obliteration may, in rare instances, be just below the valve cusps, which themselves are normal and well formed (Fig 35)

Some degree of infundibular maldevelopment is also usually present, which is not surprising considering that the pulmonary channel is functionless Occasion-



35 Photograph of example of pulmonary atresia which occurred immediately *below* the semilunar valves which are well formed life was maintained by a small persistent ductus

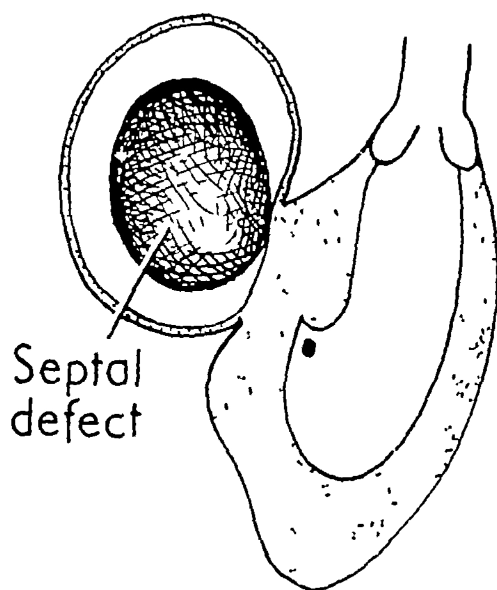
ally the right ventricle is well formed In the case depicted in Figure 35 there was tricuspid stenosis and a small thick walled infundibulum but no ventricular septal defect, the circulation in the pulmonary artery being maintained through a persistent ductus In most cases the ventricular septum is defective

The embryological fact of the fusion of the distal part of the bulbus cordis into the truncus arteriosus, and the anatomical description of the arterial root' which is formed by this distal portion (see p 21), supply the explanation of these forms of pulmonary atresia which must be due to imperfect development or to obliteration of the most distal part of the bulbus cordis

Tricuspid atresia

This type is really a group of varying degrees of imperfect development or atresia of the right ventricle and especially its inflow portion. It, in fact, illustrates admirably the twofold construction of the right ventricle, for in some cases there is almost complete suppression of the inflow portion yet the infundibulum may be almost normal. In its severer form there may be no right ventricle at all.

Characteristically the tricuspid orifice is greatly stenosed or obliterated and



36 Diagram of tricuspid atresia in which the outflow tract of the right ventricle is almost normal

the sinus or inflow portion of the ventricle does not exist or is vestigial The infundibulum may be present and moderately well developed with a small ostium connecting it to the atrophic body of the ventricle If the tricuspid orifice is completely obliterated the only route for the blood is through a persistent foramen ovale or septal defect and thence into the poorly developed right ventricle through a ventricular septal defect As the right ventricle is either non-functioning or poorly functioning it is rare to find the infundibulum completely normal and it may exist as a small thick-walled chamber Often there is a severe high infundibular stenosis or even complete pulmonary atresia, the infundibular cavity may then contain only clot If pulmonary atresia is present the ductus arteriosus is usually persistent (Fig 35) The specimen depicted diagrammatically in Figure 36 shows a well-developed infundibulum with a good-sized pulmonary artery and normal valve cusps The sinus is completely obliterated and the infundibulum opens by a small stoma into the left ventricle A large atrial septal defect is shown

CHAPTER IX

Fallot's tetralogy

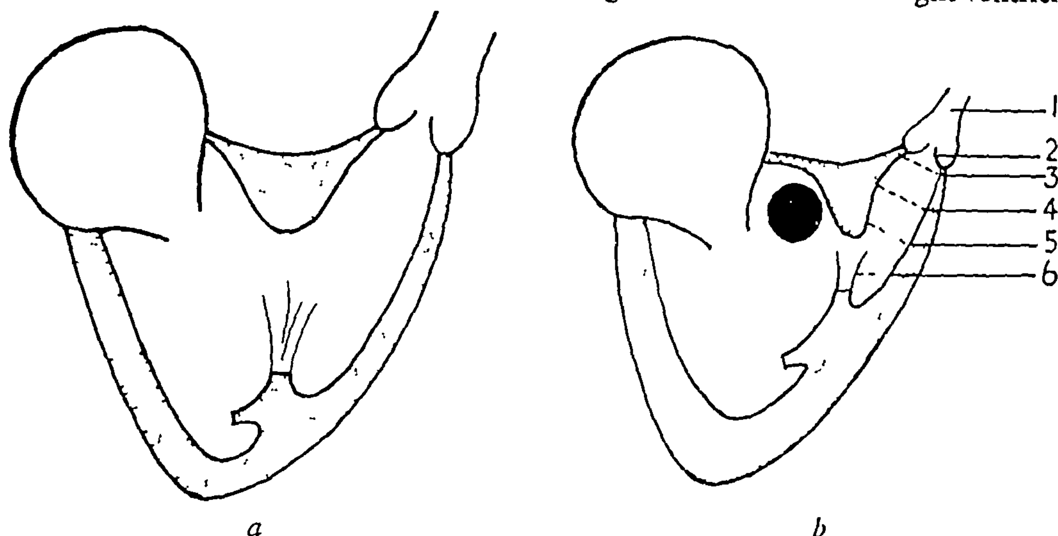
This is the most important condition to be described for it constitutes some 75 per cent of all cases of congenital cyanotic heart disease and forms the biggest group of cases of congenital pulmonary stenosis The term 'tetralogy' emphasises that the stenosis is associated with a ventricular septal defect, an over riding dextroposed aorta and a fourth element—hypertrophy of the right ventricle. This last seems a faulty presentation for the muscular hypertrophy is not a primary maldevelopment but a physiological change secondary to the pulmonary stenosis and probably also to the right to-left shunt. It is rather specious to include it as a fourth developmental abnormality

In fact, the more one studies these cases clinically and pathologically the less satisfactory does the general term 'Fallot's tetralogy' appear to be since it encourages the acceptance of a whole group of cases which, although having a number of clinical features in common, are often dissimilar anatomically

In this connection it is necessary to emphasise once more the importance of careful observation of the heart at operation, i.e in the living, functioning state, and of not being misled by what is seen at post mortem when the heart is inert, rigid, contracted and often hardened by fixation. It is commonly stated that the obstruction in Fallot's tetralogy is 'muscular' or 'tubular', the implication being that it extends over a considerable distance. I have not been able to satisfy myself that the obstruction is of this nature, it is usually annular or diaphragmatic and it would seem that most people repeating these statements have either not

examined specimens themselves or have looked at contracted specimens and have misread what they have seen It is especially easy in a fixed, contracted specimen to suppose that the obstruction is muscular because the walls of the infundibulum may be closely approximated, whereas in life an adequate, albeit diminished, lumen is present In a proportion of cases a localised hypertrophy of muscle forms the basis or foundation of the chief obstruction which is essentially fibrous It is not possible to accept the observations and conclusions of Donzelot, d'Allaines *et al* (1952) and Soulié *et al* (1952), most of the specimens used for their examinations seem to have been old, rigid post-mortem ones

Figure 37 compares the anatomical arrangement of the normal right ventricle

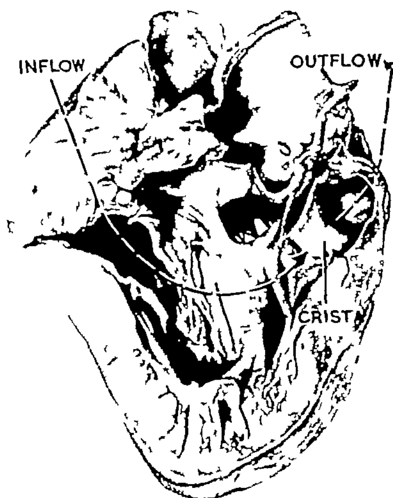


37 Diagram of the normal right ventricle (*a*) and of the right ventricle in Fallot's tetralogy (*b*), in which are indicated the various levels at which stenosis may occur

1 Pulmonary atresia
2 Valvar stenosis
3 High
4 Intermediate } infundibular stenosis
5 Low
6 Subdivision of the right ventricle

(*a*) with that seen in Fallot's tetralogy (*b*) It will be seen that there is partial suppression of the outflow tract or infundibulum In contrast, the inflow tract is larger and hypertrophied, a septal defect lies in the inflow chamber immediately behind the crista supraventricularis Although the outflow channel is small, it is nevertheless adequate to carry a sufficient blood-flow to the lungs except for an added stenosis that may occur at one or at two levels The highest level of obstruction, indicated as (1) in Figure 37, is pulmonary atresia, (2) indicates valvar stenosis and 3, 4, 5 and 6 indicate various levels of infundibular stenosis A valvar and an infundibular stenosis may co-exist

Figure 38 is a photograph of an actual specimen of Fallot's tetralogy which demonstrates these features The rather diminutive crista supraventricularis can be seen separating the small infundibulum or outflow tract from the large thick-

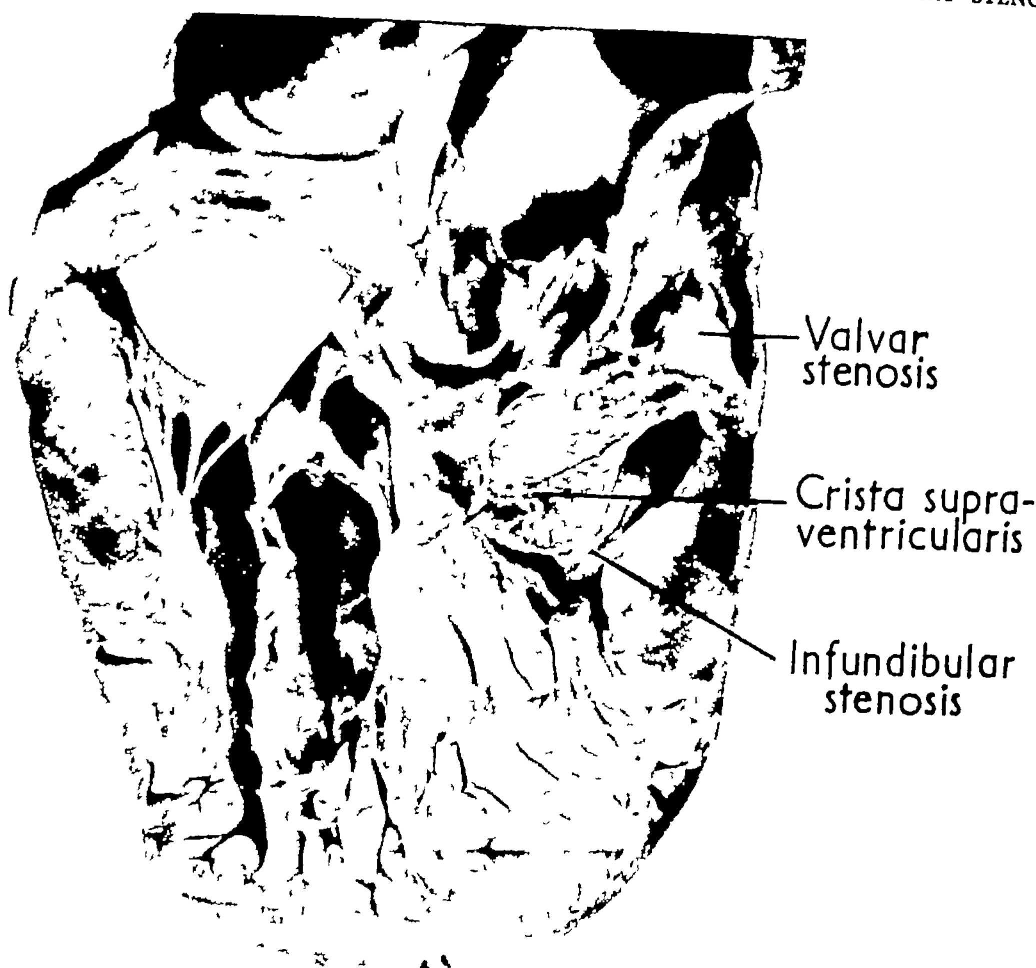


38 Photograph of specimen in Fallot's tetralogy· there is a valve stenosis

walled inflow chamber, in the anterior and upper part of which lies the septal defect. Although the outflow tract is so much under-developed, so much smaller its lumen is in fact adequate except at one level that is at the valve where there is a severe stenosis Figure 39 shows another example in which the same features can be seen except that there is a combined valvar and infundibular stenosis

In Figure 40 can be seen the formation of the infundibular canal in yet a further case of valvar stenosis occurring in Fallot's tetralogy· here again although smaller than normal, the lumen of the infundibulum is quite adequate, the true obstruction being at the pin hole valve orifice seen in the distance. The form of the crista supraventricularis is especially important in this specimen as it indicates the mode of formation of this structure from fusion of the right and left septal bands, *a* and *b* (Fig 11, p 14); the septal defect is also well displayed.

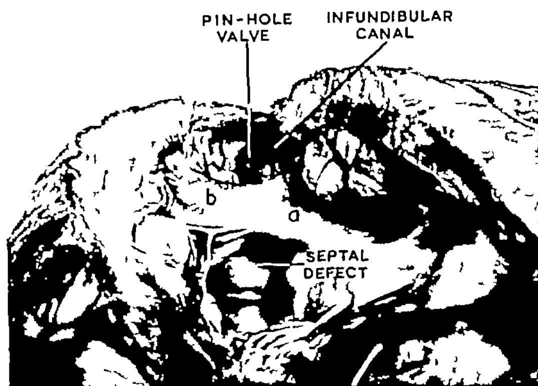
This basic anatomy and the expanded details now to be described form the bedrock on which must be founded any operative treatment for the relief of the stenosis or for total correction of the deformity



39 Photograph of specimen in Fallot's tetralogy, there is a combined valvar and infundibular stenosis

Valvar stenosis

It is surprising that a few years ago there was great opposition to acceptance of the teaching that pulmonary valvar stenosis occurs with moderate frequency in Fallot's tetralogy It would, in fact, be surprising if it did not occur when we recall that the anomaly is dependent upon malformation of the bulbus cordis, from which the valves also are developed Actually, the literature contains a number of descriptions of valvar stenosis in Fallot's tetralogy Thus Keith (1909a) in a report on 270 cases found that a valvar stenosis was present in 25 out of 63 cases (40 per cent) of what can be described as examples of Fallot's tetralogy. Brown (1939) states, 'In cases where there is an arrest of the developmental expansion of the infundibulum, the infundibulum is foreshortened and may exist as a small fusiform cavity with thickened walls In about half of the cases there is also fusion of the pulmonary cusps ' Sellors and Belcher (1950) mention that out of 65 cases examined at operation 20 were thought to have a valvar stenosis and 45 an infundibular one They do not, however, give their criteria



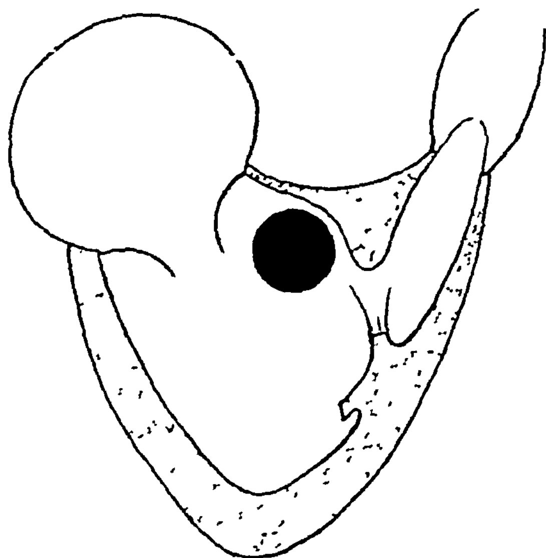
40 Photograph of specimen in Fallot's tetralogy in which can be seen the raphe of fusion of the right and left septal bands (*a* and *b*) to form the crista supraventricularis

for diagnosis nor do they state if pressure observations were made. In Abbott's series (1936) it is difficult to be sure of the exact incidence from the tables given.

My own operation experience covers 168 cases of Fallot's tetralogy in which a direct operation was performed on the pulmonary stenosis and a careful diagnosis of the type and site was made according to criteria laid down elsewhere (p 84) The distribution of the types was

	Number	Percentage
Valvar stenosis	59	35
Infundibular stenosis	72	43
Combined valvar and infundibular stenosis	37	22
Total	168	

The condition of the valvar stenosis varies and reflects the degree of hypodevelopment present. Thus at one extreme we see those cases of valvar atresia (Fig 34) or near valvar atresia in which there is a pin hole-size opening with no trace of independent cusp formation. At the other extreme the commissures of the fused triple cusps can be detected in the hemispherical diaphragmatic or domelike projection which is formed by the billowing out action of the



41 Diagram to show post-stenotic dilatation of the pulmonary trunk in Fallot's tetralogy This is uncommon

ventricular pressure (Fig 38) The fused cusps may be conical or nipple-like (Fig 39), in these examples the commissures may not be symmetrical.

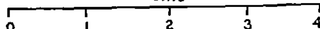
In the more severe forms the pulmonary valve is often bicuspid instead of tricuspid and the two cusps are fused along their margins, giving a small oval or round opening This type is more common with the more severe forms of infundibular hypoplasia

Where the valve is tricuspid and is better developed the stem of the pulmonary artery may show a fusiform post-stenotic dilatation with obliteration of the sinuses of Valsalva (Fig 41), although it is extremely rare to see the artery at all large and certainly almost never as big as occurs in pulmonary valve stenosis with a closed ventricular septum

The commonest type, in fact, indicates marked developmental arrest, and is of considerable interest and of great practical importance because it is easily overlooked on inspection at operation In this the fused valves form a globular or almost spherical membrane with a small central aperture (Fig 42) This globular valve distends the first part of the pulmonary artery which conforms to its shape, immediately above it there is a waistlike 'constriction' of the artery which really indicates its true small size, beyond this again is seen a post-stenotic dilatation. The whole artery is diminutive On first examination the appearances suggest the sinuses of Valsalva are present Careful inspection reveals the swelling is uniform and not trilobular, as are the sinuses, and it may then be confused with the dilatation due to a high infundibular stenosis. The spherelike structure can, however, be seen and felt to become *tense in systole* and *lax in diastole* which is diagnostic of a valvar obstruction The palpating finger can also feel the tiny emergent jet on the summit of the dome The artery beyond the valve is lax in systole, the low tension contrasting sharply with the high tension of the valve sphere The artery is almost emptied of blood in diastole, as the tense valve relaxes the small amount of blood it contains is quite enough, as it is sucked back into the ventricle, to empty almost completely the poorly-filled small distal artery.





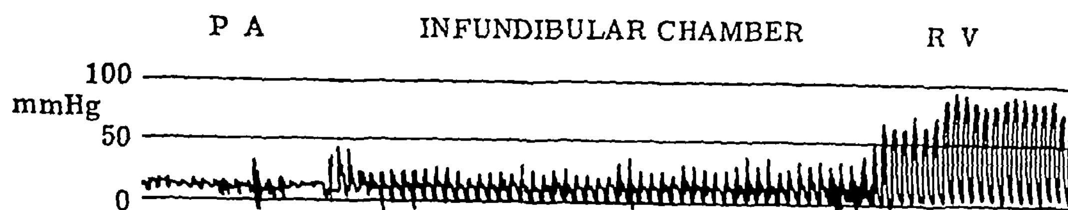


42 Globular valvar stenosis typical of Fallot's tetralogy

The small size of the pulmonary artery in valvar stenosis, and especially the constriction just distal to the valve, makes the present technique of closed valvotomy by valvotome and expanding dilator often rather ineffective; the dilating instrument may fit the lumen and it may therefore not be possible to obtain sufficient dilatation to split the valve widely An open technique is really needed when the artery is so small

It should be emphasised that the pulmonary outflow tract, including the pulmonary artery, is commonly much smaller in cases of valvar stenosis than in infundibular stenosis It seems as if valvar stenosis is associated with a greater retardation of development of the bulbus, in its extreme form it is seen as valvar atresia.

When the valvar stenosis is combined with an infundibular stenosis the fact may be recognised in most cases by external inspection and palpation of the outflow tract exposed at operation. Thus, light palpation of the valve cone will give an idea whether the full ventricular pressure is transmitted to it or whether the tension is only slightly higher than in the lax pulmonary artery, indicating that a more proximal infundibular stenosis is reducing the ventricular pressure before it can reach the valve cone. Deformation of the surface of the infundibulum, as will be described later will support the supposition of an associated infundibular stenosis. The diagnosis can be completed for certain by electromanometric pressure readings which will not only identify the presence of two changes in pressure —two stenoses—but will also indicate the relative importance of the two,



43 Electromanometric tracing of combined valvar and infundibular stenosis

whether the valvar or the infundibular stenosis is the more important or whether they play an equal part.

Figure 43 shows a combined valvar and infundibular stenosis before valvotomy and infundibular resection

It is remarkable how the presence of a true and predominant valvar obstruction can be overlooked or ignored in examination of a specimen post-mortem. Once an abnormality of the infundibulum is observed this is often taken to be the significant obstruction and no further proper search is made. In the fixed contracted state of the dead heart this might be excusable but even then a valve with an effective opening of 2–3 mm must be significantly stenosed. Examination of the living heart at operation reveals the true state of affairs with much greater accuracy if the changes mentioned above are sought and if electromanometric pressure recordings are made

The radiological diagnosis of valvar stenosis in Fallot's tetralogy and its differentiation from infundibular stenosis will be dealt with in a collective description later in this chapter

Infundibular stenosis

The basic problem in the infundibular stenosis in Fallot's tetralogy is, as already explained, an arrested expansion or development of the infundibulum so that it is smaller than normal Although in all cases the infundibulum is small, and sometimes extremely so, the functional lumen is adequate, apart from the stenosis, except in the group in which hypoplasia is extreme.

Although various 'types' of infundibular stenosis are depicted in Figure 44 and are to be described separately, it should be appreciated that fundamentally they all follow the same pattern; they are dissimilar only as a result of the level of the fibrous constriction which forms the real obstruction within the partly suppressed infundibulum Thus, in its simplest form, the obstruction is at the junction between the sinus and infundibulum (Figs 30 and 37), the usual moderate demarcation between the two parts of the ventricle is represented by a contracted muscular septum with an ostium surrounded by fibrous tissue so as to form a small rigid opening between the two parts of the ventricle and acting as an obstruction

In the other types the true obstruction lies within the infundibulum at one of the arbitrary levels depicted in Figure 37, i e it may be low, intermediate or high.

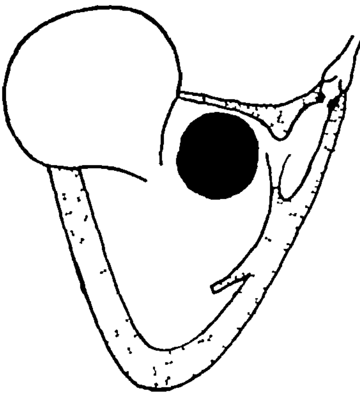
It may be only a slight annular thickening or it may be a firm raised linear fibrosis, sufficient to form a diaphragmatic type of obstruction. Providing the obstruction is not too near the pulmonary valve there will always be a post stenotic infundibular chamber varying in size with the distance of the obstruction below the valve and with the thinness of the outer wall The formation of this infundibular chamber is due to the same process of 'post-stenotic dilatation' described in connection with pulmonary valvar stenosis. When the infundibular stenosis is high the beginning of the pulmonary artery is affected by the post stenotic dilatation (Fig 44B)

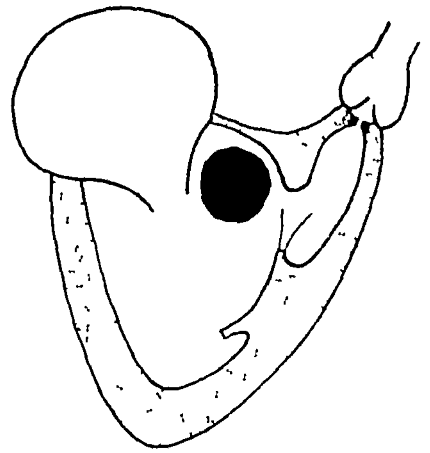
Whatever the level of the obstruction there is always a 'ground-glassing' of the endocardium of the infundibulum i.e the endocardium is thicker and whiter than normal and, according to Keith (1909), this is because the endocardial tissue is fibrocellular in texture; it is embryonic rather than inflammatory in its microscopic structure. It is now included under the term endocardial fibroelastosis

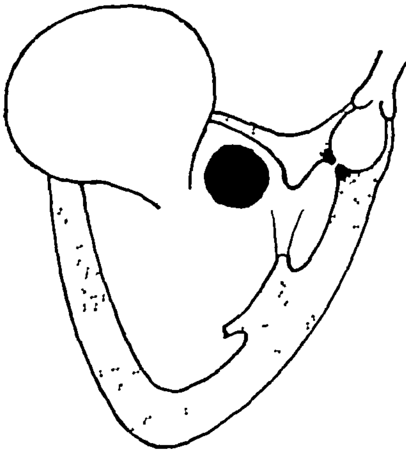
As the years pass, this endocardial thickening becomes denser and more opaque so that it may form a thick layer The greatest increase in fibrous thickening occurs at the narrowest part of the lumen, whether it be a diaphragmatic or annular narrowing in the infundibulum or at the valve itself; the daily wear and tear over the years causes increasing fibrosis amounting to a dense, hard, often cartilage-like scar and ultimately, calcification. Calcification is commonest in the valve but is also seen in infundibular stenosis The dense endocardial fibrosis may extend over the crista supraventricularis

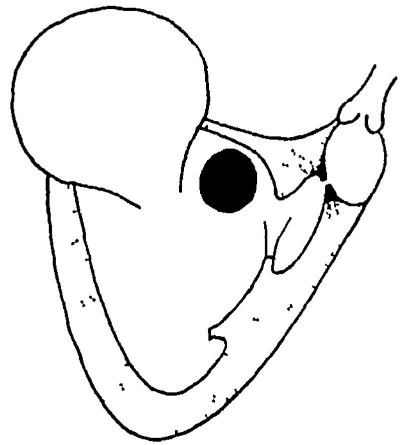
The effect of the annular band or diaphragmatic like stenosis with its endocardial thickening is to tether the outer or anterior wall of the infundibulum to the anterior surface of the crista supraventricularis which here forms the posterior wall of the infundibulum, this it does because the thickened endocardium passes from the crista to the anterior wall of the infundibulum rather like a loop or a stirrup This tethering effect can actually be observed in many cases on the outer surface of the infundibulum where the level of the stenosis may be demarcated by a furrow or groove which is deeper and more definite in systole. In the infant and child this is relatively supple but it becomes tougher and more rigid as the years pass and so its tethering action increases In this way it holds back the wall of the infundibulum from growing properly, in other words it continuously aggravates the relative backward development of the infundibular canal which cannot keep pace with the growth of the individual and of the heart as a whole. Thus an infundibular stenosis becomes *relatively* more severe as the patient gets older, and it also becomes *absolutely* more severe as a result of progressive fibrosis of the stenosed ostium, contracture of this fibrous tissue and silting up of the orifice by deposition of platelets and their organisation and the formation of warty nodules or vegetations

This process of steadily increasing stenosis is important in considering treatment, for it should be clear that the earlier treatment is carried out the better, because, with the relief of the stenosis, the infundibular canal, indeed the whole of the pulmonary channel, can then grow with the patient. If operation is delayed









D

44 Diagram of types of infundibular stenosis. A Hypoplasia B High stenosis C Intermediate D. Low

until growth has ceased there is a limit to what can be achieved in satisfactory relief of an infundibular obstruction.

This of course only applies to operations involving direct relief of the stenosis itself; the indirect or anastomotic operations do nothing to encourage the growth and development of the infundibulum, in fact by short-circuiting it they favour its silting up

The maldevelopment of the infundibulum inevitably affects its muscular construction the crista supraventricularis is always smaller and less developed than normal (Fig 38) and is sometimes very small in the more severe forms Figure 40 shows how the primary formation of the crista by fusion of the right and left bulbar ridges (p 14) may remain evident, pointing to partial arrest of development. In the same way the right and left extensions of the crista down the lateral and septal walls to the apex are smaller and less well developed

The imperfect development of the musculature is often also seen in the outer or anterior wall of the infundibulum distal to the stenosis Inevitably this part of the muscle of the right ventricle will not thicken as does the muscle proximal to the stenosis, but it may still form a substantial layer More often, however it is thin and may contain muscle only just above the stenosis, and this rapidly diminishes until the greater part of the outer wall is devoid of muscle and presents as a thin walled bulge on the surface of the ventricle through which the dark blood is plainly seen.

This thinning of the outer wall of the infundibulum in association with the process of post-stenotic dilatation, can result in quite a large and prominent infundibular chamber being formed when the stenosis is low The projection of this chamber on the surface is a notable diagnostic feature both radiologically and at operation.

Although it is emphasised that the true obstruction in infundibular stenosis lies dominantly at one level, even though there is some arrest of the development of the infundibulum as a whole, there is no doubt that the effect of this stenosis is aggravated by the systolic contraction of the infundibulum. In fact it is probable that the efficiency of emptying is, as it were, cut short before the height of systole as the walls of the narrowed infundibulum come together This has been demonstrated by Hilario Lind and Wegelius (1954) using a rapid biplane angiocardiograph technique. Rodbard and Shaffer (1955) have demonstrated the same by cardiac catheterisation (see p 102) These matters will be discussed more fully later when the control mechanisms of the right ventricle are being considered

Having presented these general observations on infundibular stenosis we may now proceed to the individual types (Fig 44).

(i) Hypoplasia of the infundibulum

This type (Figs 44A and 45) is due to almost complete arrest in the development and expansion of the infundibulum so that it is only a narrow muscular channel, too small to carry an adequate blood flow to the lungs, even so it is still



45 Hypoplasia of infundibulum

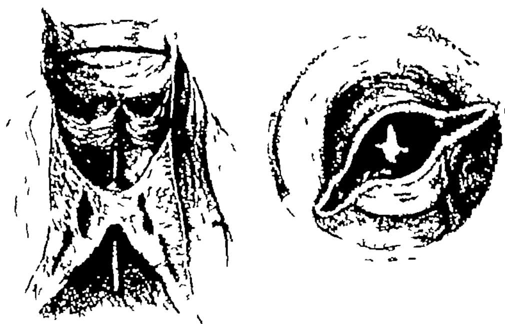
further narrowed at one level by a secondary fibrous stenosis or by valvar stenosis, as always occurs in Fallot's tetralogy The pulmonary artery is small and poorly developed The condition in its severest forms is only little removed from pulmonary atresia and is inevitably most often seen in infants and young children Inevitably, because in them any degree of hypoplasia of the infundibulum must be more severe because of the small size of their outflow tract and also because early death is common and fewer live to be seen at an older age

The narrow channel surrounded by muscle is likely to be grossly contracted by rigor mortis, and even more so if fixed in formalin, so that the appearance of a purely muscular obstruction is aggravated

It is, in a sense, true that the obstruction in this type is 'muscular' in that the whole infundibular canal is inadequate If the child lives a few years the infundibulum will grow sufficiently with the rest of the heart to lessen, to a certain extent, the extreme smallness of the lumen seen in the infant and it is then easier to appreciate the part played by the localised fibrous constriction in completing the obstruction of the poorly developed outflow tract (Fig 45)

(ii) High infundibular stenosis

In this type (Fig. 44B) the degree of arrest of development is not so extreme as in type A, the infundibular part of the ventricle is larger and the pulmonary artery is better developed although often still too small. The obstruction is a firm annular or diaphragmatic thickening of the endocardium just below the valve level. The condition is admirably portrayed by Peacock (Fig 46) and is also shown in Figures 47 and 48 In older patients widespread secondary fibrosis occurs at the obstruction, and may extend proximally over the walls of the imperfectly developed infundibulum to form a thick layer of lining (endocardial



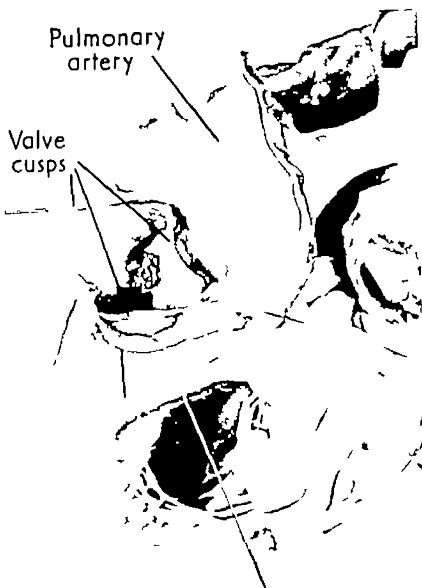
46 High infundibular stenosis as portrayed by Peacock.

fibroelastosis) over the surface of the crista supraventricularis Figure 47 also shows considerable fibroelastosis.

As the stenotic ring is very near the valves there is no space for a true chamber to form from stenotic dilatation, but the effect of this process causes the valve ring and the first part of the artery to dilate in a spherical manner, thus mimicking the post-stenotic dilatation seen in valvar stenosis The appearance of the artery is rather like that of a drum-stick; distal to the globular formation the walls of the vessel are parallel and the tension of both the dilatation and the trunk is the same and is low; this is in contrast to valvar stenosis in which the proximal dilatation is tense, and immediately beyond it is a constriction before the more distal dilatation. The post-stenotic dilatation of the pulmonary artery may extend to its main branches, especially the left one.



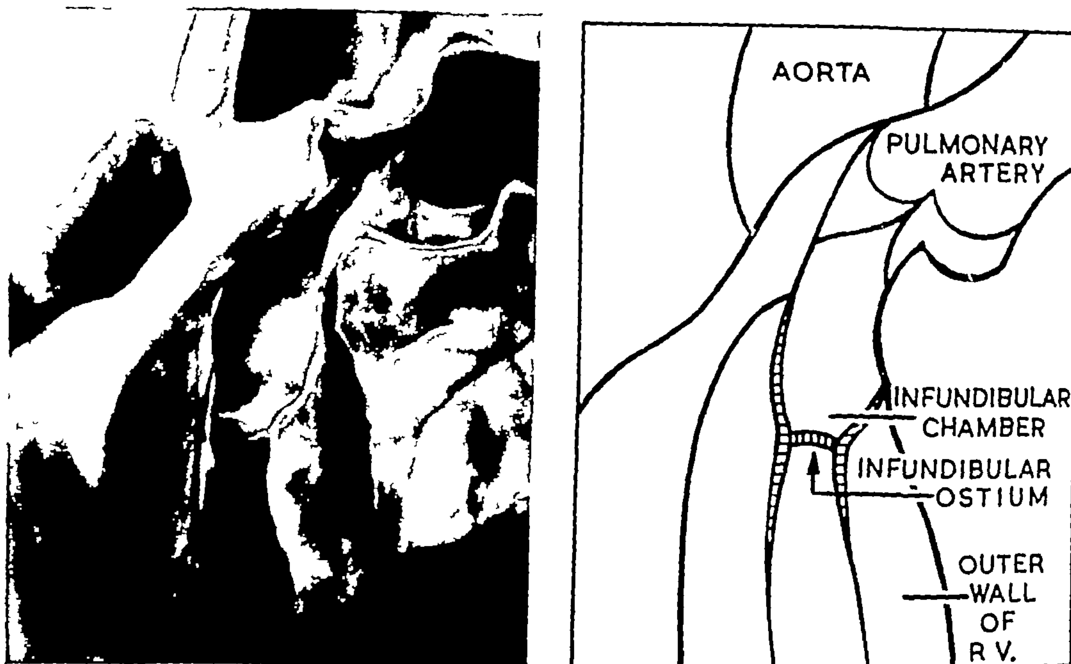
47 High infundibular stenosis



48 High infundibular stenosis

(iii) *Intermediate infundibular stenosis*

In this type (Figs. 44C and 49) the actual fibrous obstruction is situated from 1 cm to 2 5 cm. below the valve level so that a definite post-stenotic infundibular chamber forms This chamber is lined by thickened endocardium and is limited proximally by a fibrous obstruction either annular or diaphragmatic (Fig 49) The amount of fibrous tissue in the obstructing ring or diaphragm varies, as a general principle the younger the patient the less fibrosis there is, in older patients a dense, fibrous, rigid, often verrucose margin surrounds the narrowed entry into the chamber The outer wall of the chamber may be thin and fibrous, or partly or wholly muscular although always much thinner than the wall of the ventricle proximal to the obstruction. When the wall is deficient in muscle a



49 Intermediate infundibular stenosis

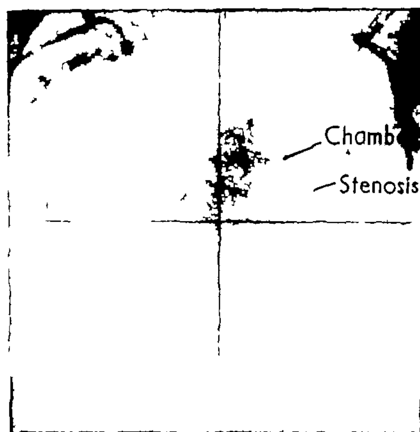
globular swelling, dark blue from the contained blood, projects from the surface of the right ventricle below the pulmonary valve. This can be observed on plain radiography and can be further demonstrated by angiocardiograms (Figs 50, 55, 56 and 57) Figure 51 shows the stenotic ring removed by infundibular punch resection

(iv) Low infundibular stenosis

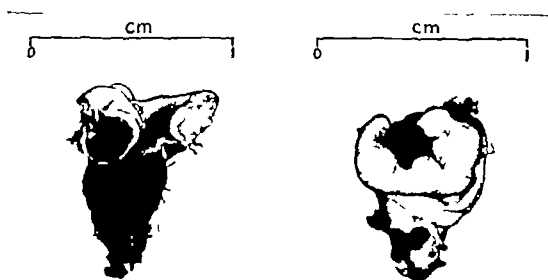
This type (Fig 44D) and type 'C,' the intermediate, are really only variants, being arbitrarily divided by the level of the fibrous obstruction below the valves, and thus in the size of the infundibular chamber which varies between 2 5 cm and 5 cm in length If the outer wall of the chamber is uniformly muscular it will not project markedly, but if the wall is thin and the muscle extends into it for only 1–2 cm. the chamber is large and prominent and can be demonstrated radiologically as well as at operation (Fig 31, p 44)

Muscle forms the basis of the obstructing ring, but the margins of the ostium are always fibrous Sometimes two or even three openings may be present.

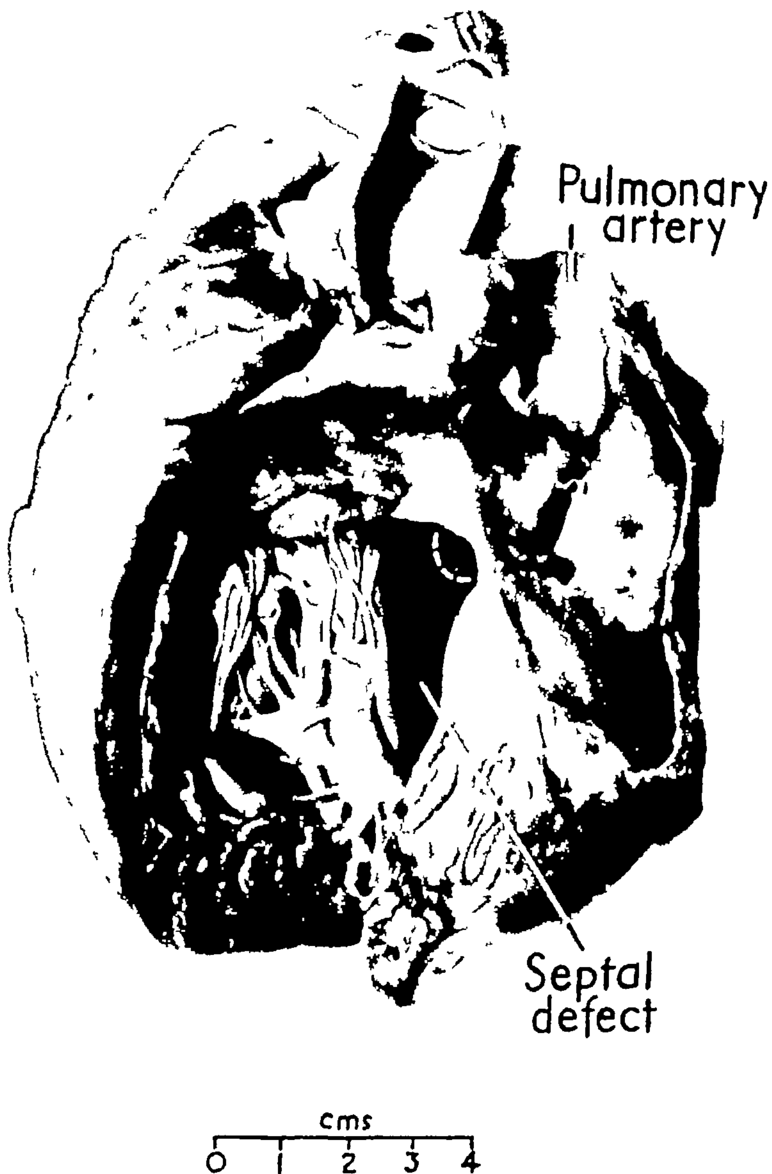
The specimen shown in Figure 52 is an example of an unusually large infundibular chamber in association with a low stenosis In fact, from the size of this chamber it resembles closely the type seen in pure infundibular stenosis described earlier It has a large muscular septum situated at the junction of the sinus and infundibulum and perforated by an ostium with fibrous margins, its outer wall thins out so that it is almost completely deficient in the upper part, indicating the development of the infundibulum was definitely defective.



50 Angiocardiogram of intermediate infundibular stenosis



51 Photograph of specimen obtained by punch resection from patient in Fig 50

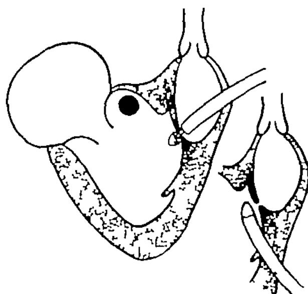


52 Low infundibular stenosis, note the large septal defect and the large infundibular chamber with a thin outer wall

However, there is also a large ventricular septal defect measuring some 4 cm in diameter, so large as to throw the two ventricles virtually into one

An important practical feature in infundibular stenosis is the obliquity of the septum between the infundibular chamber and the main body of the ventricle This obliquity is slight or absent in a high stenosis, but becomes quite marked in low stenosis, in fact in its lower part this septum lies nearly parallel to the outer wall of the ventricle with only a narrow space intervening The stenosed opening between the two compartments is higher up on the partition in such a way as to make surgical access difficult Figure 53 illustrates the problem diagrammatic-

53 Diagram to show how the obliquity of the secondary septum in low infundibular stenosis may make punch resection from below unfavourable.



ally in the usual technique of punch resection of the infundibular stenosis the punch is inserted through the ventricular wall below the obstruction, it will be seen at what a disadvantage it may lie. If it is introduced from above, it approaches the stenosis and the partition much more favourably This technique can only be used if the infundibular chamber is large

Combined valvar and infundibular stenosis

Since the deformity in Fallot's tetralogy is due to maldevelopment of the bulbus cordis which extends distally to include the pulmonary valves as well as forming the infundibulum, it follows that malformation of the valves is to be expected in combination with infundibular stenosis as well as an independent condition

In the table on page 55 which gives an analysis of 168 cases treated by direct operation, a combined valvar and infundibular stenosis was present in 37 (22 per cent)

The valvar stenosis has already been fully described the infundibular stenosis is usually of high intermediate type. I cannot recall seeing a low infundibular stenosis combined with a valvar stenosis, doubtless this is because a really low infundibular stenosis represents a less severe malformation whereas a combined valvar and infundibular stenosis is ordinarily a more severe malformation

The presence of two stenoses may be recognised before operation by angiocardiography or by cardiac catheterisation, or by both. At operation the combined features of the two lesions are to be recognised. The final diagnosis comes from electromanometric pressure recordings made with catheter and needle puncture; in this way, in addition to confirming the presence of the double lesion, the level of the infundibular stenosis and the relative importance of the two stenoses can be assessed (Fig 43 p 58)

The septal defect

The defect in the septum is situated immediately behind the crista supraventricularis and its lower margin and anterior and posterior boundaries are rounded and muscular (Fig 38), its upper margin is the aortic ring although it has no real upper margin as it opens directly into the aorta

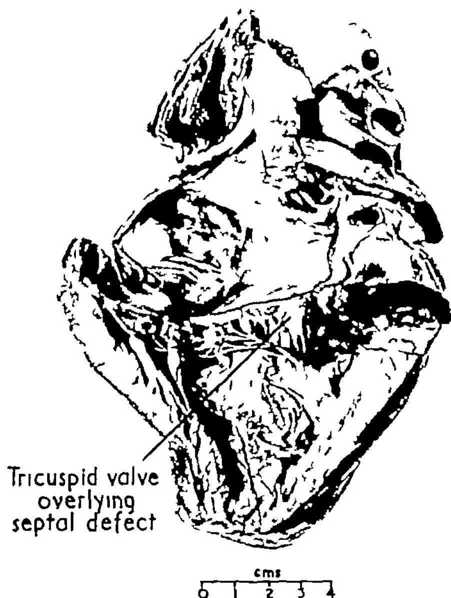
The defect is usually smallest when the infundibulum is well formed, this rule is not invariable for in the specimen shown in Figure 52 in which the obstruction is very low, there is a huge ventricular septal defect 4 cm wide

The anatomy of the septal defect has become of greater importance now that direct operation for its closure is possible Although it appears, especially in the dead heart, to be a rigid round or oval defect, its upper margin can be very easily approximated to the aortic margin before fixation, and even the fact of the overriding aorta does not prevent this In the extreme forms of dextroposition the pulmonary artery is usually considerably hypoplastic, and closure of the septal defect would then be unwise because the poor pulmonary channel would not be able to carry the extra circulation effectively for the heart to readjust itself to such profound alteration of its mechanics Although there is usually no true upper margin to the defect, the actual aortic ring provides strong and suitable material for suturing It is also worth remembering that the right atrium is a direct anterior relation of the aorta adjacent to the defect and sutures could in fact be passed through this to help to close the upper septal margin

An important posterior relation of the defect is the septal cusp of the tricuspid valve with its tendons and papillary muscle In Fallot's tetralogy the septal defect is very close to the cusp and may extend under it, but rarely to the same degree as may occur in isolated ventricular septal defect in which the septal cusp may cover quite an appreciable amount of the defect. Figure 54 shows a specimen of Fallot's tetralogy in which the defect is covered by the tricuspid valve

When the defect is small its posterior margin reaches no further back than the atrioventricular rings, and sutures inserted for closure can pass directly from the muscular wall of the defect to the aortic root When the defect is larger it extends further backwards and in so doing comes to lie more and more under the atrioventricular rings, i e. it extends beneath the mitral and tricuspid valves which form part of the roof, the septal cusp of the tricuspid covering its right aspect In such cases it would be wrong to attempt to close the posterior part of the defect by suturing to the aortic ring, this part must be closed by picking up with the sutures the part of the atrioventricular ring which gives common origin to the tricuspid valve on one side and the mitral valve on the other After the posterior part has been closed in this way, the more anterior part can be closed by sutures which pick up the aortic root

Another structure with an important relationship to the septal defect is the atrioventricular bundle The right branch has already been mentioned as demarcating the junction between the sinus and infundibulum of the right ventricle. The main bundle bifurcates immediately behind the posterior limit of



54 Heart in Fallot's tetralogy to show that the ventricular septal defect may be covered by the septal cusp of the tricuspid valve

the defect and the right branch runs down the septum a fraction of a centimetre in front of the posterior margin, it is well behind the crista supraventricularis

The degree of dextroposition of the aorta or over riding varies from case to case, but is again usually dependent on the degree of arrest in development of the infundibulum. In the severe forms with a high stenosis and a large septal defect the aorta seems to over ride at least 50 per cent In addition in these severe cases the ascending aorta is usually very dilated, a fact observed and commented upon by Peacock. In Chapter XI (p 87) the significance of dextroposition is more fully discussed, and especially the views of Lillehei *et al* (1955) who have indicated that it is apparent rather than real

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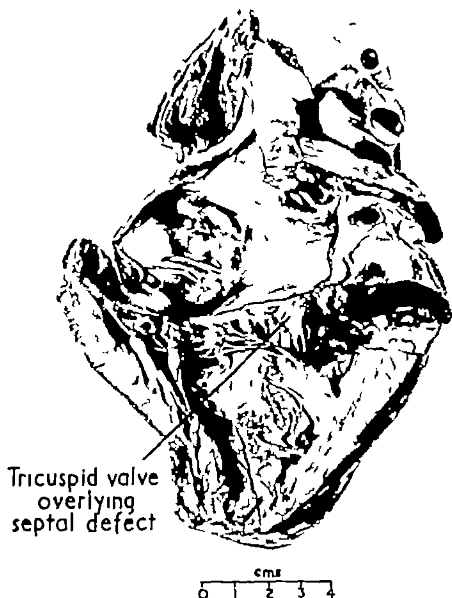
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CHAPTER X

The recognition and differential diagnosis of the type and level of the obstruction in Fallot's tetralogy

MANY of the details of the recognition of the type and level of the various forms of pulmonary obstruction emerge from a consideration of what has already been said The subject is, however, so important that it is desirable to correlate the various features and to enlarge upon them by a more detailed consideration of some of the radiological and other aspects

One of the easiest pre-operative radiological diagnoses can be made in the case of a large infundibular chamber associated with a low stenosis. Figure 55 shows clearly how the chamber stands out from the left margin of the heart shadow in the routine postero-anterior view, in the past this shadow has often been loosely described as being due to a 'prominent conus region', it is characteristic of an infundibular chamber and it is brought into even greater prominence in an oblique view (Fig 55*b*), angiocardiograms again demonstrate its presence conclusively (Fig 55*c*)

Figure 56 shows another example of a large infundibular chamber In both these cases the diagnosis was confirmed at operation and the intervening fibro-muscular diaphragm was punched away so as to relieve the stenosis, confirming that the obstruction was diaphragmatic and not long and tubular.

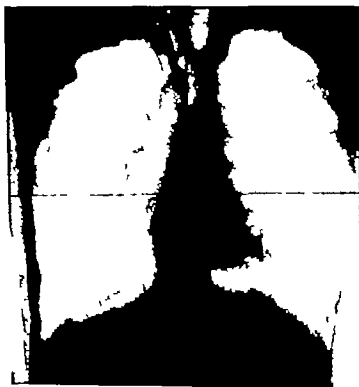
That confusion and misunderstanding exist about what is really a simple state of affairs is shown by Figure 57 As will be seen from the plain radiograph on the left, '*a*', a characteristic bulge due to an infundibular chamber is present, '*b*' is a



a



b



c

55 To show the bulge on the heart contour formed by an infundibular chamber

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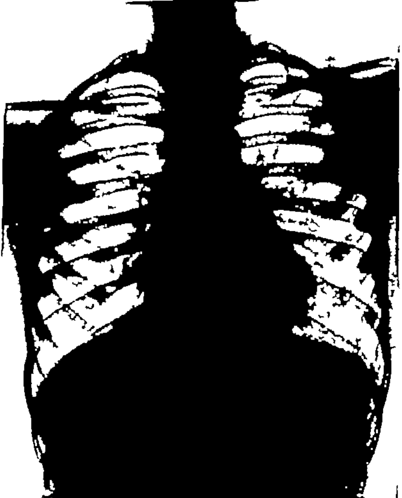


c

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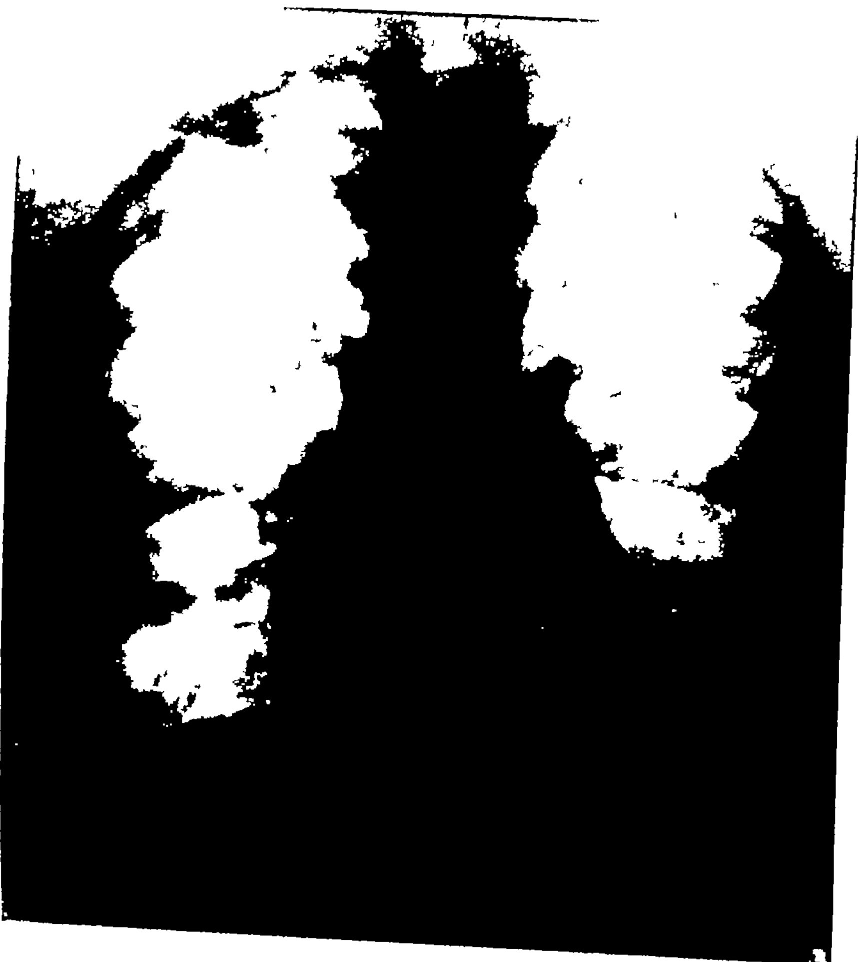


a

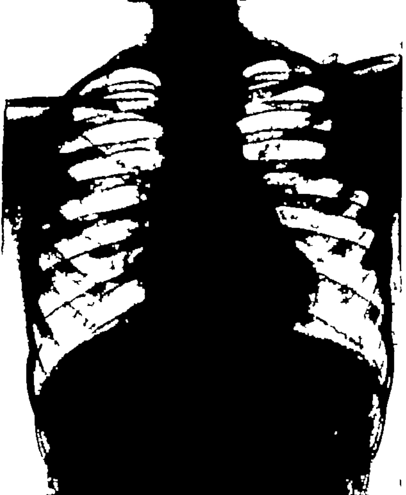


b

57 Erroneous interpretation of pressure changes observed in a case of infundibular stenosis with post stenotic chamber' incorrectly assumed to be a long narrow stenosis



56 To show the bulge formed by an infundibular chamber.



a



b

57 Erroneous interpretation of pressure changes observed in a case of infundibular stenosis with post stenotic chamber incorrectly assumed to be a long narrow stenosis.

radiograph taken at cardiac catheterisation in which two crosses have been marked, the top one shows the site of pressure change at the valve level and the lower one the site of change from an intermediate pressure to the high pressure of the right ventricle proper. Between the two crosses the pressure was constant at 19/10 mm Hg and it was assumed by those concerned that this represented a long tubular type of obstruction which would be very difficult to treat by direct operation The problem was referred to me for comment

Obviously there is no evidence at all for a long stenosis, the pressure changes merely mark the low tension to be expected as the catheter tip traverses the infundibular chamber so clearly shown in '*a*', the abrupt rise at the lower x marks the diaphragmatic-like stenosis before the body of the right ventricle is reached This error of interpretation is common and arises from the preconceived idea, almost superstition, which is engendered by the teaching that the stenosis of the infundibulum is typically long and tubular The falsity, of course, melts away before reasoned observation of the true anatomical state and of simple thought on the true nature of the pressure changes

That the catheter can be of value in localising the level of the stenosis is shown by Figure 58, in which the catheter tip is seen in '*a*' above the pressure change and in '*b*' below it, that this lies in the infundibulum and not at the valve is clear

Figure 59 shows an electromanometric tracing obtained at operation from a proven case of infundibular stenosis and in the upper tracing can be seen the abrupt rise of pressure as the catheter enters the right ventricle. In this tracing, taken before the stenosis was resected, the pressure in the pulmonary artery is so low that it does not emphasise that the low diastolic pressure in the infundibulum is equal to that in the ventricle This is, however, clearly shown in the lower tracing made after infundibular resection

It is also well shown in Figure 43 in the tracing made at operation on a case of combined valvar and infundibular stenosis, the pulmonary artery pressure is not much lower than the pressure in the infundibular chamber but the drop in diastolic pressure marks the transition from artery to infundibulum, the greater pressure change as the ventricle is entered indicates that the infundibular stenosis is the more important

It must be emphasised that pressure observations must be made at operation to confirm the site and severity of the obstruction even though this may appear obvious from external examination of the heart, certainly the presence of two obstructions can be confidently elucidated only in this way Moreover, the effect produced by the operation on the stenosis or stenoses must be assessed at operation by further measurements and tracings

Just as an infundibular chamber should be readily recognised, so can a valvar stenosis when the artery shows post-stenotic dilatation This applies only with certainty to 'pure' pulmonary valve stenosis in which the artery can usually be seen on the plain radiograph (Fig 27, p 39), and shows up well in the angiocardiograph as diagrammatised in Figure 60A, it will be remembered that when

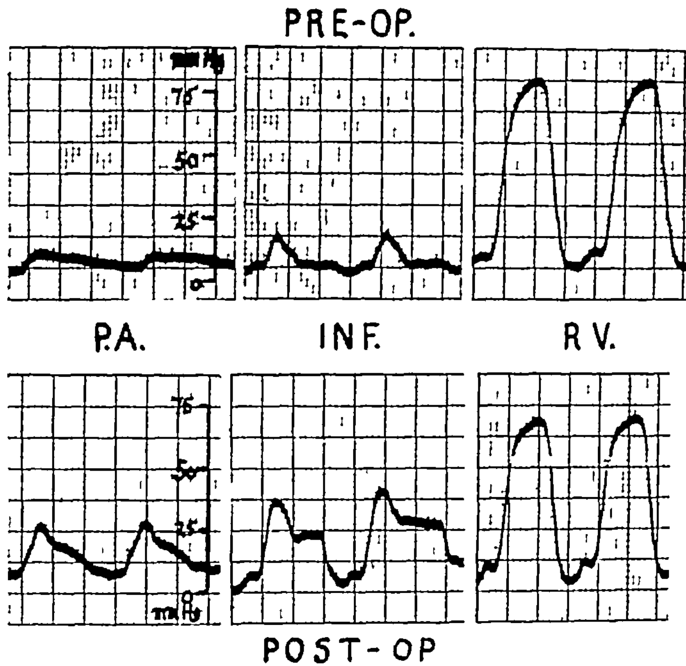


a



b

58 Use of cardiac catheter to localise the site of stenosis



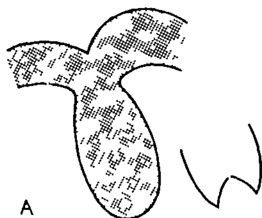
59 Electromanometric tracing of infundibular stenosis

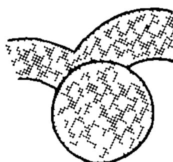
the heart is very large the artery is pushed up so as to be horizontal and is therefore seen end on or foreshortened (Fig 28), this also is shown in Figure 60B

I have only once seen in Fallot's tetralogy sufficient dilatation of the stem of the pulmonary trunk on a plain radiograph to indicate a valvar stenosis. A moderate or minor bulge is sometimes seen near the origin of the artery, but never enough to differentiate from a high infundibular stenosis

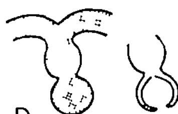
If the type of valvar stenosis is as depicted in Figure 41, that is with a general moderate dilatation of the artery, the angiocardiograph may reveal a general ovoid dilatation (Fig 60C) similar to that seen in 'pure' pulmonary valvar stenosis, although the artery is much smaller As a similar dilatation may be caused by a high infundibular stenosis it is important to try and assess whether the length of the dilated segment suggests that only the pulmonary artery is affected or whether it extends below the valve level, thus including the highest part of the infundibulum It may be difficult to decide this, especially as the amount of foreshortening of the artery varies with the amount it is pushed upwards by right ventricular hypertrophy It is then helpful to consider the shape of the proximal part of the opacity, a smoothly rounded end suggests a valvar stenosis and a pointed or conical end suggests a high infundibular stenosis (Figs 60D and E) Figure 61 is an angiocardiograph of an undoubted case of high infundibular stenosis with much general post-stenotic dilatation of the pulmonary artery *and of its left branch* This is also shown in diagram in Figure 60F, the dilated left pulmonary artery may be visible as a 'comma' on the plain radiograph It can occur in either valvar or high infundibular stenosis and so cannot be used as a diagnostic feature

In Figure 62 a smooth globular termination is seen in a proven case of high



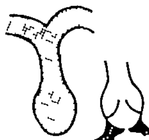


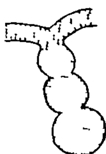
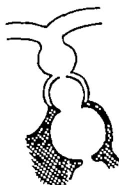




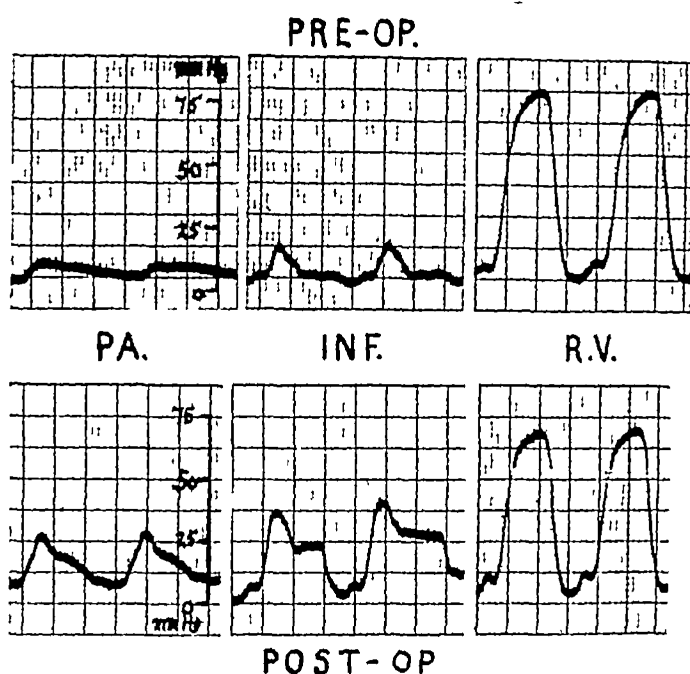






60 Diagram to show interpretation of various angiocardiographic appearances. (For full description see text.)



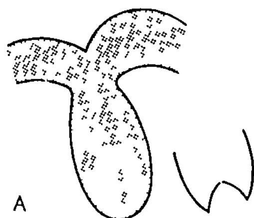
59 Electromanometric tracing of infundibular stenosis

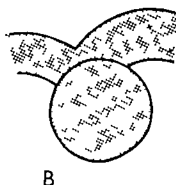
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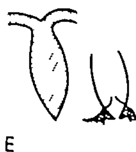
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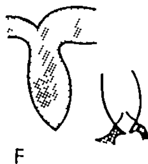


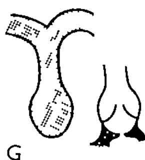


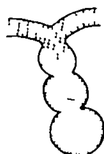
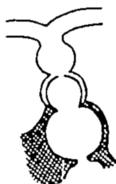










60 Diagram to show interpretation of various angiocardiographic appearances. (For full description see text.)

61 High infundibular stenosis with large left pulmonary artery



62 High infundibular stenosis with rounded proximal end



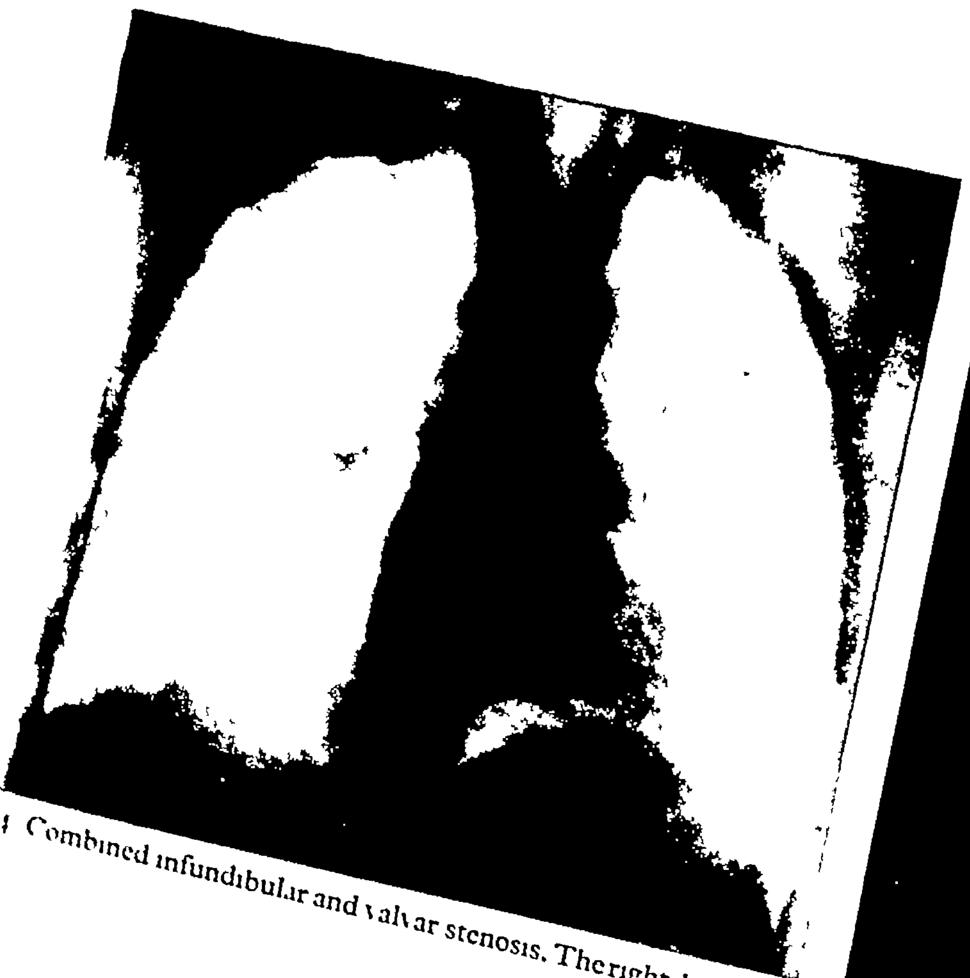
63 High infundibular stenosis.

infundibular stenosis but the slight extra length indicates this condition although it would be difficult to exclude a valvar stenosis with certainty before operation (Fig 60G) In Figure 63 the length of the pulmonary artery is sufficient to diagnose with fair confidence that the proximal bulge is due to a small infundibular chamber; this is also suggested by the uniform density of the pulmonary artery

Figure 64 shows a combined valvar and high infundibular stenosis, the bilobed or cottage loaf appearance of the proximal shadow suggests this and is supported by the slight but definite break in filling of the pulmonary trunk immediately distal to the smaller upper part of the swelling (Fig. 60H)

A lateral view may help greatly in estimating any extension below the valve level because in this view foreshortening is corrected. With an ordinary venous angiocardiograph the shadow of the superior vena cava may be superimposed on that of the pulmonary outflow tract and it is, therefore, better to use a selective technique in which the contrast medium is injected via a catheter in the right ventricle. Figure 65 shows such a radiograph of a valvar stenosis in which it is even possible to identify the linear filling defect due to the valve itself and also the emergent jet. Jönsson *et al* (1953) show some beautiful illustrations of selective angiocardiographic demonstrations of the types of pulmonary stenosis

The globular type of valvar stenosis is the commonest in Fallot's tetralogy and has already been described and has been depicted in Figure 42 It is shown



61 Combined infundibular and valvar stenosis. The right-hand figure shows an enlarged view of the opacified stenotic region, *cf* Fig 60H.



65 Selective angiocardiogram to show valvar stenosis

diagrammatically in Figure 60D, instead of an ovoid dilatation of the whole length of the main pulmonary artery there is only a globular swelling at its base. Another feature that may point to a valvar stenosis is persistence of the shadow within the valve globe or cone after the pulmonary artery has emptied, this is, of course, a reflection of the obstruction present.

At operation, recognition of a valvar stenosis may be easy and the criteria already laid down for pure pulmonary valvar stenosis apply equally The most crucial test is actual palpation, either with one finger or between finger and thumb, of the dome of the valve becoming hard and tense in systole. The greatest difficulty exists when a small globular valve is present; at first glance the localised

dilatation resembles persistence of the sinuses of Valsalva, perhaps somewhat smoothed out and suggesting dilatation distal to a high infundibular stenosis Careful palpation should, however, reveal that the stem of the pulmonary artery is under low tension all the time, *whereas the globular first part becomes tense in* systole and relaxes in diastole The reverse is the case if the sinuses of Valsalva are present, they seem to relax in systole and become slightly more tense in diastole. In addition certain other features of a high infundibular stenosis are absent.

The bulge of a globular valvar swelling may often be seen and felt through the intact pericardium, but so can the bulge due to a high infundibular stenosis and it is impossible to differentiate the two unless the pericardium is opened Then the bulge of a high infundibular stenosis is seen to extend definitely below the level of the valve ring Usually it appears to be half above and half below the ring, as is shown by the presence of a thinned-out layer of muscle fibres on its surface, becoming rapidly thicker proximally where a definite depressed groove appears on the surface of the ventricle in systole This groove indicates the tethering of the outer wall of the infundibulum to the crista supraventricularis by the linear fibrosis which constitutes the maximum obstruction Moreover, although the sinuses of Valsalva are involved in the dilatation it is still possible to recognise their presence and structure. *Palpation fails to identify the tense dome of the* stenosed valve in systole, and although a jet-like thrill can be felt, it is not quite the same as with a valvar stenosis; it is usually not so fine and precise, its origin is less easily determined and *it can be felt below the valve level if a finger is placed on the proximal part of the bulge, especially at the level of the constriction* Even when the outer wall of the small chamber is thick enough not to form a bulge, light palpation will reveal that the wall is thinner than that of the main body of the ventricle and in this way the actual line of transition can be identified In doubtful cases the transition can be found by electromanometric readings with a needle by making punctures on each side and narrowing down the gap, a catheter can also be used for securing a withdrawal tracing and the exact position of the stenosis identified by measurement of the length of catheter within the heart when the pressure change occurs

The same technique may have to be used when there is a large infundibular chamber associated with a low stenosis but concealed because of good muscular development of its outer wall, when the outer wall is thin there is, of course, no difficulty

Finally, if any doubt remains, a finger can be inserted through an incision low down in the right ventricle and the exact site of the stenosis identified

It has been objected that the pattern of infundibular stenosis does not follow the descriptions given here, that such a description is erroneous in affirming that the form of the stenosis is eminently suitable for operative resection except in a small proportion of cases in which there is extreme hypoplasia of the outflow tract

The success of operations for the direct relief of pulmonary stenosis must rest

on the correctness of the surgical anatomy given above. My experience of direct operative relief of the stenosis in cases of Fallot's tetralogy makes me quite confident of the correctness of these observations, observations made on the living, functioning heart. Analysis of the actual results of the direct operations on the stenosis shows good or very good results in some 75 per cent of cases (Brock, 1955) Such a high proportion of good results would be impossible unless the operations were well founded anatomically

CHAPTER XI

Forms of transposition; common ventricle

THIS chapter deals with a rather varied group of conditions in which pulmonary stenosis plays a part but is usually not the most important anomaly.

Aorta and pulmonary artery both arising from right ventricle

This is a good example of the inadequacy, which was mentioned earlier, of the term 'Fallot's tetralogy'. Clinically and radiologically a patient with this malformation may appear to be a typical case of the tetralogy, but the actual malformation demands separate classification Both ventricles are present, but the aorta arises wholly from the right ventricle, which also gives rise to the pulmonary artery Blood can leave the left ventricle only through a ventricular septal defect to enter the right ventricle and thence to the two great vessels There are examples of lesser severity in which the aorta arises perhaps 90 per cent from the right ventricle and over-rides the left ventricle perhaps 10 per cent Some blood can, therefore, pass directly into the aorta from the left ventricle, but the greater part has to pass through the septal defect It is difficult to say at what degree of over-riding a diagnosis of Fallot's tetralogy becomes unacceptable Arbitrarily, it might be said that anything more than 75 per cent over-riding of the septum moves the case into a separate group

This is because such a large degree of over-riding clearly indicates a dominant

origin of the aorta in connection with the right ventricle; in the typical Fallot's tetralogy the aorta unquestionably arises primarily from the left ventricle, and indeed many think that the septal overlap is not real but is apparent or illusory and results from the loss of fixation of that part of the aortic ring which has lost its support due to the septal defect. In fact, if one excises this portion of the ventricular septum in a normal heart, it creates the impression of an over riding aorta if viewed from the right ventricle (Lillehei *et al*, 1955) In these cases there is no difficulty in correcting the apparent dextroposition by suturing the margin of the septal defect to the aortic ring In cases of genuine total or subtotal transposition of the aorta in which it arises dominantly from the right ventricle, it would be impossible to do such a repair to separate the outflow tracts of the two ventricles In fact if repair were possible it would, in contrast, result in total separation of the aorta from the left ventricle.

Total dextroposition of the aorta certainly demands separate classification, the higher degrees of dextroposition in which the aorta arises almost entirely from the right ventricle would appear to belong rather to this group than to a straightforward Fallot's tetralogy Certainly the prognosis is worse, both without operation and also after operation in which the immediate mechanical derangement may be profound and even fatal

The stenosis may be infundibular but has been valvar in a sufficiently large number of cases to suggest that it may be the commoner lesion in this type. The valvar obstruction may be of such high degree that it constitutes almost an atresia.

The aorta and pulmonary artery in these cases may lie side by side like a two-barrelled gun, the aorta may then represent the right reptilian aorta arising from the recess behind the crista supraventricularis which has already been mentioned as representing the outflow tract of the reptilian right aorta (p 7)

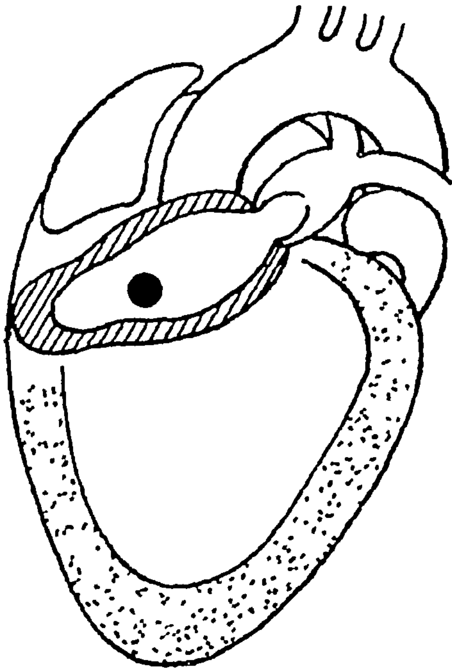
Transposition of the great vessels with pulmonary stenosis

It is no part of this monograph to give a full account of the development and anatomy of transposition of the great vessels, for this complex anomaly may exist without any form of pulmonary stenosis

However it may also be accompanied by a pulmonary stenosis which is usually valvar but may be infundibular The pulmonary artery lies directly behind the aorta and is largely hidden both radiologically at operation and anatomically

Common ventricle (fusion of inflow portion of right ventricle with left ventricle)

Various forms of common ventricle may be met with in which there is also a pulmonary stenosis, but in the type selected here for special mention the leading feature is absence of a ventricular septum so that the infundibulum exists as a small annexe of a large common ventricle (Fig 66) There is no trace of the inflow



66 Diagram to show common ventricle with a small, partly separate, right ventricular outflow tract, a low infundibular stenosis is seen.

portion of the right ventricle The infundibulum is always small and connects with the common ventricle by a small ostium which is similar to, perhaps identical with, the stenosed stoma usually seen in infundibular stenosis between the infundibulum and the body of the right ventricle. In addition there is usually an abnormality of the pulmonary outlet, either a high infundibular stenosis or a valvar stenosis, this may be of the nature of a valvar atresia.

This type must represent an early and severe abnormality of the bulbar fusion for not only is fusion incomplete but expansion of the bulbus is greatly retarded In one case, of a child aged 8 with moderate cyanosis and disability, the infundibulum, viewed at operation, was raised and quite distinct from the common ventricular mass so as to form a small muscular channel or tube about 0 75 cm wide and 2–3 cm long, just like a diminutive and separate bulbus cordis; clearly a high-grade atavism There was only a tiny proximal stoma connecting with the ventricle and distally a high-grade valvar stenosis amounting almost to atresia Life was supported by a small persistent ductus.

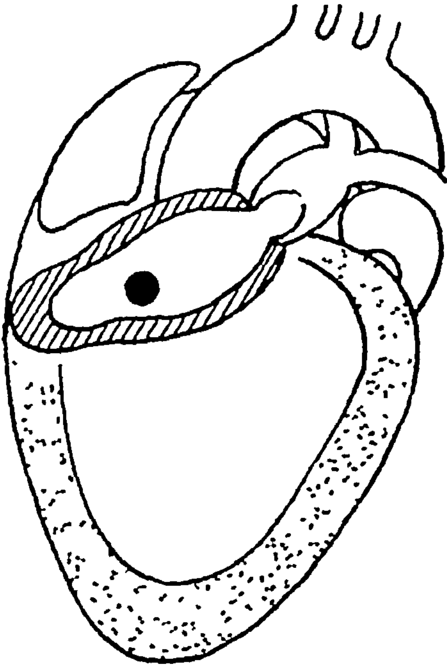
CHAPTER XII

Summary Direct operations for pulmonary stenosis, physiological changes due to direct operations

Investigation of the morbid anatomy of pulmonary stenosis reveals that the obstruction is almost always due to maldevelopment of the bulbus cordis, the distal part of which forms the first part of the pulmonary artery including the pulmonary valves, the proximal part is incorporated within the right ventricle to form the infundibulum. Stenosis may occur at various levels, but, conveniently, can be considered as affecting either the distal or the proximal part of the bulbus When the distal part is at fault, atresia of some portion of the pulmonary artery occurs, or pulmonary valve atresia or pulmonary valve stenosis Pure' pulmonary valvar stenosis with an intact ventricular septum is also due to malformation of the distal part of the bulbus

Malformation of the proximal part of the bulbus gives rise to infundibular stenosis Combined valvar and infundibular stenoses imply that both the proximal and distal portions of the bulbus are at fault.

Although the infundibulum exhibits various grades of under-development, in every case the maximal obstruction is localised, being annular or diaphragmatic; the narrowest part of the channel may exhibit muscular thickening in its base, but the actual stenosis or ostium is surrounded by fibrous tissue. In older patients this becomes progressively thicker, warty and even calcified. The normal systolic contraction may add a functional infundibular stenosis or may aggravate an existing anatomical one.



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In one type (hypoplastic) the whole of the infundibulum is atrophic and the pulmonary channel is totally inadequate, although even in this type there is a secondary zone of maximal fibrous stenosis or there is a valvar obstruction

Where the infundibulum is not so hypoplastic it is convenient to classify the types on the basis of the level of the maximal obstruction This may be 'high', i e immediately below the valves, no infundibular chamber is formed. At other levels it may be 'intermediate' or 'low', in all of these a post-stenotic dilatation occurs giving rise to an infundibular chamber which may be prominent externally if its outer wall is deficient in muscle It is imperative to recognise that in all types, although the infundibulum may be so much smaller than normal as to give a reduced pulmonary channel, this is usually adequate and the effective obstruction is always annular or diaphragmatic In the simplest and rarest form the obstruction is at the junction between the body and the infundibulum of the ventricle, the cavity of which is then virtually separated into two parts by a muscular septum perforated by an ostium with a rigid fibrous margin In this type there may be no ventricular septal defect, in all the others a septal defect exists

In a proportion of cases of Fallot's tetralogy, probably between 30 and 40 per cent, the effective obstruction is not infundibular, in spite of the infundibulum being small, but is valvar In some 20 per cent an infundibular and a valvar obstruction co-exist

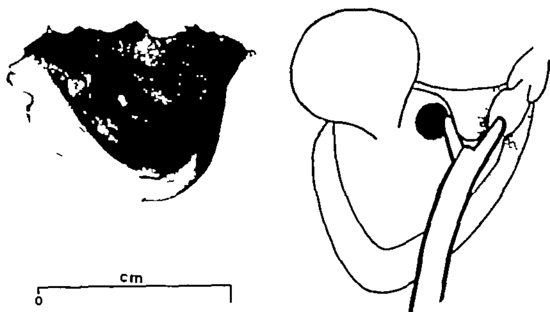
These observations on the nature of the actual obstruction are fundamental in considering operative treatment

The direct operative relief of pulmonary stenosis

This is no place to debate the relative values of the indirect anastomotic operations and direct relief of the obstruction itself, this has been discussed elsewhere (Brock, 1954, 1955) It is sufficient to point out that if the actual obstruction can be relieved the heart is directly benefited and the mechanics of the circulation are not further complicated by an artificial ductus arteriosus.

In 'pure' valvar and 'pure' infundibular pulmonary stenosis there can be no doubt that valvotomy or infundibular resection are the operations of choice If the obstruction is not relieved and a short-circuiting operation done, no relief is given to the burdened right ventricle and an additional strain is thrown on the left ventricle The already overtaxed right heart has to cope with a further increase of circulating blood and sooner or later fails

Direct relief of the obstruction, whether it be valvar, infundibular or the two combined, is also feasible in a very high proportion of cases of Fallot's tetralogy. The anatomical basis for these procedures has been set out in the preceding chapters and summarised above and its truth has been abundantly confirmed during the experience personally gained in 168 direct operations on Fallot's tetralogy during the last 7 years. That the anatomical basis is soundly conceived is also proven by the high percentage (some 75 per cent) of good or excellent



67 Resection of a segment of crista supraventricularis by means of a spur punch. This, by disrupting the sphincteric like action of the infundibular muscle may destroy the control mechanism

results obtained The only lesions for which a direct operation is not feasible are those of tricuspid atresia pulmonary atresia and severe degrees of pulmonary outflow hypoplasia, especially in infants and young children.

When the crista supraventricularis is involved in the stenosis the question arises of resecting part of it to increase the lumen. This is possible as a closed procedure by using a punch forceps with a terminal bite (Fig 67) Whether it is good surgery to divide this important muscular structure is perhaps doubtful, it can be very effective in relieving the infundibular stenosis

When it is being resected two structures are vulnerable; one is the septal cusp of the aortic valve which lies above; the other is the right branch of the atrio-ventricular bundle This runs down the septum from the posterior part of the septal defect well behind the crista and would be damaged only if an instrument were directed well backwards It has never been damaged on the numerous occasions I have performed punch resection of the crista

Direct operations on the stenosis are better done at an early age so that the pulmonary outflow tract can grow with the patient. Development must be progressively severely arrested if the stenosis is not relieved, for in addition to the stenosis becoming *relatively* more severe as the patient grows older, it becomes *absolutely* more severe as a result of daily wear and tear on the stenosed passage. Ultimately the obstruction may become complete or so nearly complete as to be virtually so

Valvotomy allows progressive dilatation of the main pulmonary artery and of its branches, infundibular resection, by correcting the tethering of the walls of the infundibulum by the fibrous ring which loops out from the crista

supraventricularis, allows growth and expansion of the channel as the rest of the heart grows In other words, normal growth and progressive improvement are favoured

In patients in the late teens and twenties, in addition to permanent arrested growth of the infundibulum, secondary narrowing has occurred from progressive fibrosis Direct operation is less favourable in these older patients when the stenosis is high, it is more likely to be favourable when the stenosis is low because post-stenotic dilatation has produced a good-sized infundibular chamber. The only disadvantage is that if the muscular wall of the distal part of this chamber is thin and atrophic it may stretch under the influence of the increased pressure and flow, and secondary functional pulmonary regurgitation may develop (see Chapter XIII).

That increase of size of the pulmonary artery does in fact follow a direct operation has been observed in several cases when a second operation has been performed Thus, in a young boy aged 5 years the pulmonary artery was less than 1 cm in diameter in association with a valvar stenosis Valvotomy was followed by a good result for 2 years and then cyanosis and disability began to return At a second operation 5 years after the first one the pulmonary artery was found to be of normal size, the valvar stenosis had been corrected, an infundibular stenosis was present and was successfully resected

This is an important observation for it means that even though the pulmonary outflow tract is severely under-developed, it can be restored to full growth by a successful direct operation It should then be possible at a second operation, should it be necessary, to close the ventricular septal defect without fear that the pulmonary channel could not carry the additional flow of blood if the stenosis is totally removed This must always be a fear in certain cases of Fallot's tetralogy now that technical developments allow simultaneous resection of the stenosis and closure of the septal defect Although a proportion of cases will be primarily suitable for such a one-stage definitive curative procedure, there will always be a proportion in which the pulmonary stenosis will have to be relieved as a first stage and the septal defect closed as a second stage

It has been suggested that the simple direct operation for the relief of valvar or infundibular stenosis in Fallot's tetralogy is incomplete and therefore obsolescent It is doubtful, in the light of the above observations, if this is correct

Correction of the right-to-left shunt

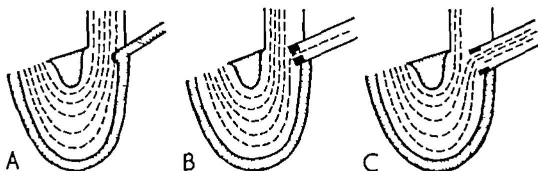
In addition to lessening cyanosis and disability by increasing the blood-flow to the lungs through the normal channel, direct operations on the obstruction act beneficially by another mechanism

Part of the cyanosis and disability is due to deficient blood-flow to the lungs, part is due to the right-to-left shunt through the septal defect In pulmonary valvar stenosis with a persistent foramen ovale there can be no question at all of this latter mechanism because cyanosis may appear only after many years and can

be corrected completely by successful valvotomy In malformations of the Fallot's tetralogy type in which there is a ventricular septal defect, a similar effect is produced (Fig 68) Thus, if there is considerable obstruction to the outflow into the pulmonary artery, a large part of the output of the right ventricle inevitably passes into the aorta with considerable mixing of venous and arterial blood. If the obstruction to the pulmonary artery, whether valvar or infundibular, is relieved, a greater part of the output of the right ventricle can pass to the lungs and a *correspondingly smaller amount is diverted into the aorta*; the arteriovenous mixing is, therefore, reduced.

If the optimum relief of the pulmonary stenosis is obtained by operation the right to-left shunt will be completely abolished, cyanosis and disability will be completely relieved.

In some cases the shunt is not only corrected, it may be reversed so that it



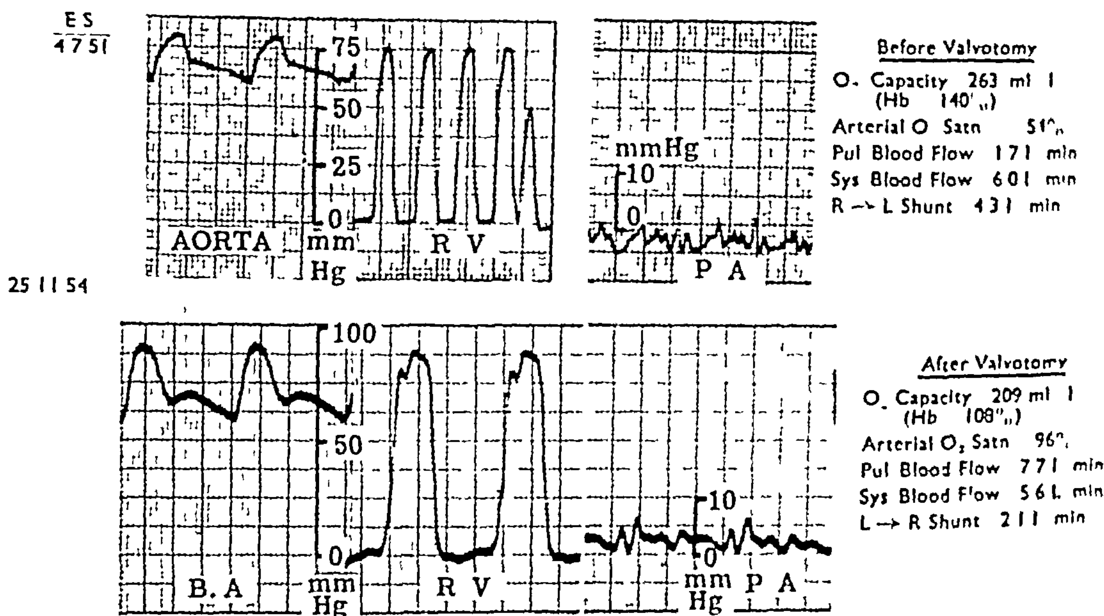
68 Diagram to show the effect of direct relief of the pulmonary stenosis in Fallot's tetralogy in correcting the right to-left shunt in addition to improving the pulmonary blood flow A. Pulmonary atresia total shunting B Fallot's tetralogy, partial shunting C. After operation, lessened shunting

becomes left to-right; the pulmonary flow being slightly greater than the systemic flow In occasional cases we have observed considerable reversal of the shunt so that the pulmonary flow becomes too great. In some of these the pulmonary artery pressure has been slightly raised as a result of the increased flow, not from increased resistance. Thus in one case the oxygen saturation rose from 86 per cent to 98 per cent, the pulmonary flow increased from a half to double the systemic flow the pulmonary artery pressure rose from 14/7 mm. Hg to 38/12 mm. Hg

Far from a severe rise in pulmonary artery pressure following direct relief of the pulmonary stenosis the rise in pressure is more often disappointingly small, indeed it may rise so little that the clinical result would appear to be inevitably unsatisfactory In fact, with even a trivial rise in pressure in the pulmonary artery the clinical result may be excellent, demonstrating that it is pulmonary *flow* rather than *pressure* that matters In many such cases the peripheral pulmonary resistance is so low that no pressure rise occurs The following case serves as a perfect example of this

The patient was a young man aged 19 years and as bad a case of Fallot's

tetralogy as it is possible to meet. He was quite incapacitated and was wheeled into the out-patient clinic in a chair He was so cyanosed, distressed and ill that it was necessary to admit him to hospital at once In due course he underwent operation and a pulmonary valvotomy was done Figure 69 shows the catheter studies in July 1951 before operation and in November 1954 after operation If the pressure curves are examined it will be found that there is but little change, the right ventricular pressure is 75 mm Hg before and about 90 mm Hg after operation, the pulmonary artery pressure was rather below zero and was between



69 Catheter studies before and after successful pulmonary valvotomy in Fallot's tetralogy (For description and discussion see text)

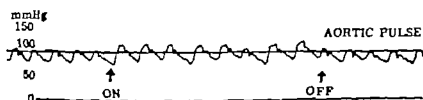
zero and 5 mm Hg after operation The clinical result, however, was as perfect as one could wish All cyanosis and disability were relieved, he gained some 3 stone in weight and from being a complete invalid is now able to do a full day's work This improvement is reflected in the other catheter findings given in Figure 69 Thus the arterial oxygen saturation has risen from 54 per cent to 95 per cent (haemoglobin has fallen from 140 per cent to 108 per cent), more striking still is that the pulmonary blood-flow has risen from 1 7 litres per minute to 7 7 litres per minute, in fact the right-to-left shunt of 4 3 litres per minute has been changed to a left-to-right shunt of 2 1 litres per minute It is difficult to imagine a more striking example of the important part played by the peripheral pulmonary resistance in influencing blood-flow.

It is possible that in later years we may observe an increase in peripheral pulmonary resistance accompanied by a rise in pulmonary artery pressure and a diminution of pulmonary flow, perhaps even reversal of pulmonary flow and recurrence of the right-to-left shunt and cyanosis This, however, remains for the future In the meantime the prospect of routine successful closure of the

ventricular septal defect under direct vision in these cases (Brock, 1954) makes it probable that the onset of such secondary changes may be entirely prevented.

Moreover the ability to close the ventricular septal defect will entirely remove anxieties and limitations in regard to too generous relief of the pulmonary stenosis The need to leave some protection for the pulmonary circulation will no longer remain and the stenosis can be dealt with as radically as possible under the same direct vision.

Arising further out of this question of alteration of the right-to-left shunt after successful operation for the relief of the pulmonary stenotic element in Fallot's tetralogy (valvotomy or infundibular resection), interesting immediate secondary physiological changes may be observed Thus, on one occasion at an operation in which infundibular resection had been completed, the anaesthetist remarked that the peripheral blood pressure had fallen and the pulse was poor; at first this was thought surprising because an excellent pulmonary blood flow could be felt as judged by palpation of the pressure and flow in the pulmonary



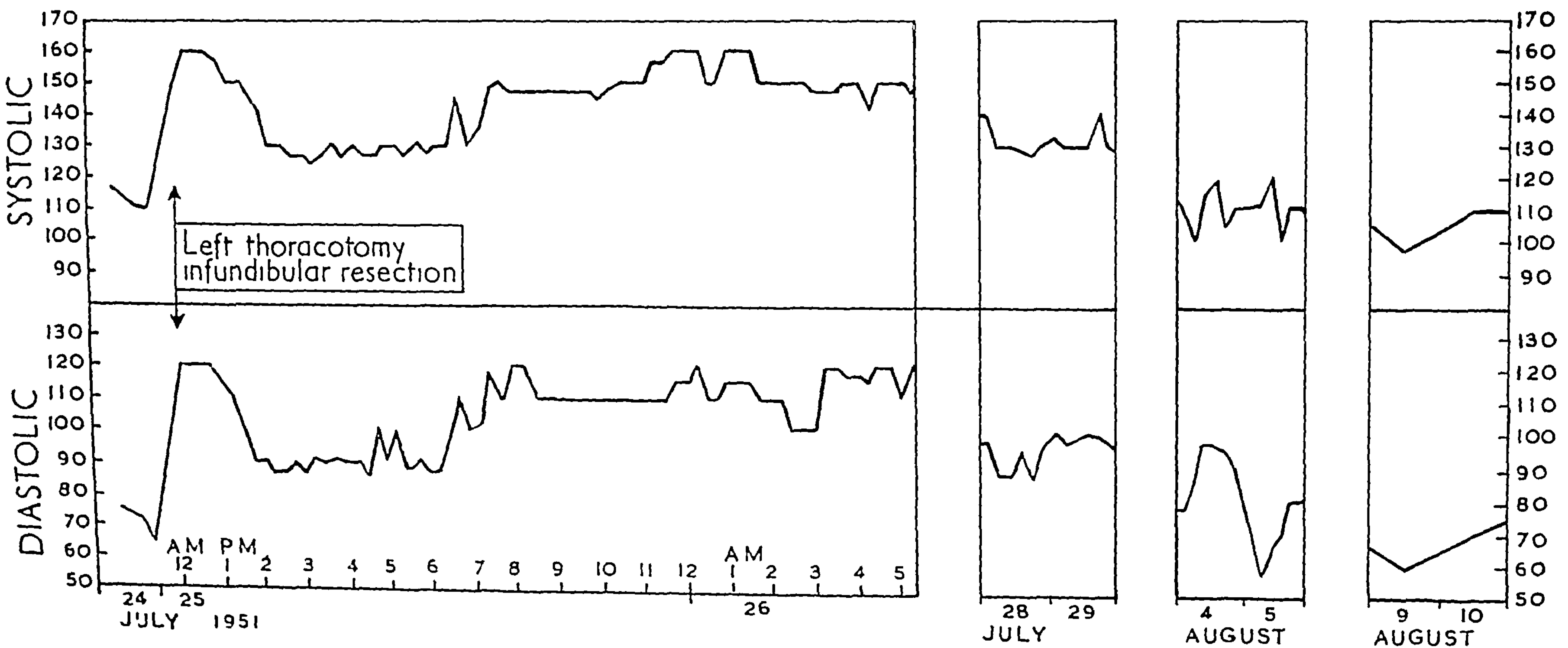
70 Aortic blood pressure tracing in a case of Fallot's tetralogy after infundibular resection When the pulmonary outflow tract was compressed the aortic pressure rose 10 mm. Hg and fell again when the pressure was released thus demonstrating variations in the right to-left shunt.

artery The more usual observation is that deterioration in the patient's general condition is accompanied by diminution in the pulmonary flow, i.e there is general depression of the circulation. In contrast in this case there was selective depression of the systemic circulation.

This suggested that the increased pulmonary flow was at the expense of the systemic output, and this was confirmed quite simply by observing the aortic and radial artery pressure while a finger was pressed on the pulmonary outflow tract and then relaxed. Figure 70 shows the effect clearly in an electromanometric tracing

This mechanism is intelligible if one accepts that the combined ventricular output cannot immediately increase to compensate for its redistribution, thus if the pulmonary flow increases the systemic flow must decrease by about the same amount. The only way the ventricles can correct this state is by increasing their output and this may be difficult in the early stages after their efficiency has been impaired by a surgical intervention

This is doubtless also the explanation of another phenomenon observable, especially in young children, during the first few days after operation, that is the occurrence of systemic hypertension. Soon after operation the patient may show evidence of greatly diminished peripheral blood flow as seen by pallor, some



71 Blood-pressure chart after a direct operation on a patient with Fallot's tetralogy showing the temporary rise in blood-pressure often observed

cyanosis and poor pulse volume; in contrast the blood pressure is high and may reach 200 mm Hg systolic or even higher This is a remarkable occurrence in a young patient who has just undergone a major ventricular cardiotomy; it would be not unreasonable to expect *hypotension* rather than *hypertension* In one child the systolic pressure 24 hours after operation rose as high as 270 mm Hg, a figure which caused considerable alarm as to its effects on the heart and concern as to whether some gross accident had occurred

The explanation seems to lie in an exaggerated response to the redistribution of the ventricular output as mentioned above the only way the ventricles could themselves correct the diminished systemic flow would be to increase the stroke volume. This can scarcely be done so soon after ventricular cardiotomy Compensation is achieved by another mechanism—peripheral vasoconstriction, this can help to maintain an adequate coronary flow while cutting down the less important peripheral circulation. The peripheral constriction is obvious clinically

Figure 71 shows the blood pressure chart in such a case; the phase of secondary hypertension and then gradual recession to normal pressure is shown. This secondary rise is best seen in young children but may be observed at all ages.

In addition to the physiological and clinical interest of this phenomenon there is an important practical bearing After cardiotomy for pulmonary stenosis with a ventricular septal defect (Fallot's tetralogy) it would be easy to be content with a normal or subnormal blood pressure; it could well be thought that this was to be expected. Actually this should give grounds for alarm, for it would indicate that the circulation is not being readjusted satisfactorily Unless there is no doubt that the patient's condition is quite satisfactory it is better to take steps to raise the blood pressure, i.e by transfusion, posture and, if necessary vasoconstrictor drugs

CHAPTER XIII

Control mechanisms in the outflow tract of the right ventricle

THE function of the right ventricular outflow tract is somewhat more complicated than is generally thought Indeed, it is doubtful if much thought in general is given to it, the usual conception is that of a simple muscular canal which contracts to aid the expulsion of the blood it receives in diastole In actual fact, the function is more sensitive, more labile, than this and is particularly concerned with a mechanism exerting a control on the pulmonary blood-flow

Although this is a function in the normal heart it assumes greater importance and is more readily demonstrable in the abnormal heart, especially when there is some form of pulmonary stenosis

The anatomy of the normal right ventricle has been described (Chapter III), and Figure 16 (p 19) illustrates the way the infundibulum stands out in a manner which shows it is anatomically partly separate from the rest of the right ventricular muscle mass and which suggests it may also be functionally somewhat separate.

This arrangement of the infundibulum was fully explained by simple reference to its development It will be recalled (Chapter I) that in the fish and the amphibian heart the bulbus cordis exists as a separate part or chamber which is not seen in the mammalian heart, in which it loses its identity by becoming absorbed into the musculature of the outflow tract of the right ventricle

The function of the infundibulum

Seeing that structure is related to function, the obvious anatomical difference between the infundibulum and the rest of the right ventricle suggests that this may be related to different function. To Keith (1904c) belongs the credit of having drawn attention to this He states, 'The great development of the bulbus arteriosus in gill-breathing vertebrates is due to the peculiar nature of their circulation. The heart is primarily a respiratory pump, the ventricle throws its load into the short aorta leading to the gills To be able to receive the ventricular load and to force it onwards, that vessel is endowed with enormous elasticity so that no valves within it could be mechanically competent were they not furnished with a muscular or elastic support, such as is provided by the bulbus arteriosus. The musculature of the bulbus, like that of the aorta, is always in action but most so in *diastole* of the ventricle. In every way the pulmonary artery resembles the short gill aorta of fishes of which it is the functional representative in air breathing vertebrates Keith then goes on to suggest that in man the pulmonary valves are rendered competent only because they are reinforced by the musculature of the ventricle which extends up to them. In support of this he quotes some experiments carried out by Gibson (1898) in which he showed that the pulmonary valves allow regurgitation if they are exposed to the weight of more than a certain column of fluid This regurgitation could be controlled by constricting the pulmonary artery by tying a cord round it exactly at the attachment of the valves Gibson felt that this showed clearly that the relative incompetence of the valves was caused by distension of the elastic artery Tested in the same way the aortic valves never allowed any escape These observations led Gibson to suggest that a mechanism of readjustment to prevent regurgitation is desirable in the pulmonary circulation and Keith supports this view He feels 'the fibres of the bulbus arteriosus in lower vertebrates are not truly discharging but regulating fibres that maintain their tonicity when the true fibres of the ventricle have passed into a state of diastole when the right heart is distended the circular fibres round the base of the pulmonary artery form a functional stricture at the commencement of ventricular systole between the infundibulum and greatly dilated artery and are a factor that must be considered in explaining the production of pulmonary systolic bruits

I have repeated these experiments of Gibson and have been able largely to verify his findings and certainly to verify his conclusions The heart from a freshly killed dog was used and long lengths of glass tubing were tied into the aorta and pulmonary artery and then filled with water The pulmonary valves soon become incompetent and permit regurgitation. The aortic valves support a much higher column but they too become incompetent; the findings thus differ in this respect from those of Gibson who stated that the aortic valve never allowed any escape. The relative efficiency of the two systems is shown by the simultaneous height of the water columns above each valve; in one experiment the aortic column was 90 cm. and the pulmonary column was 20 cm When a flat tape was tightened



72 Photograph of a dog's heart in which a column of water in a glass tube tied in the pulmonary artery is causing gross distension of the pulmonary valve ring and of the musculature of the infundibulum

round the pulmonary valve ring the onset of regurgitation could be greatly delayed, as shown by Gibson If the tape is released when the valves have been supporting a high column of water in this way, the weakness and inability of the pulmonary ring and the infundibular musculature to support the valves is strikingly seen, gross dilatation of the sinuses of Valsalva and of the pulmonary ring is seen at once (Fig 72)

I have also carried the observations a stage further In an anaesthetised dog the heart was exposed by a trans-sternal incision and the roots of the great vessels identified One needle was then inserted into the pulmonary artery and one into the right ventricle so that simultaneous electromanometric pressure recordings were made. The pulmonary artery was then partly occluded by a clamp so that the pressure rose, regurgitation at the pulmonary valve occurred rapidly, thus with an initial pulmonary artery pressure of 16/14 mm Hg regurgitation occurred when the pressure reached 26 mm Hg systolic In contrast, when the same observation was made on the aorta regurgitation did not occur until the systolic pressure was over 300 mm Hg Thus the initial aortic pressure was 98 81 mm. Hg, there was no regurgitation at 261/180 mm Hg, regurgitation began at 316/163 mm Hg and was more severe at 320 mm. Hg.

Attempts were made to show that the onset of pulmonary regurgitation could be retarded when the tonus and power of the right ventricle was enhanced by calcium chloride or by adrenaline, although this appeared to be the case the results of the experiment were not entirely conclusive, adrenaline seemed cer-

tainly to delay the onset of regurgitation. (I am indebted to Dr P Fleming for assistance in making these observations)

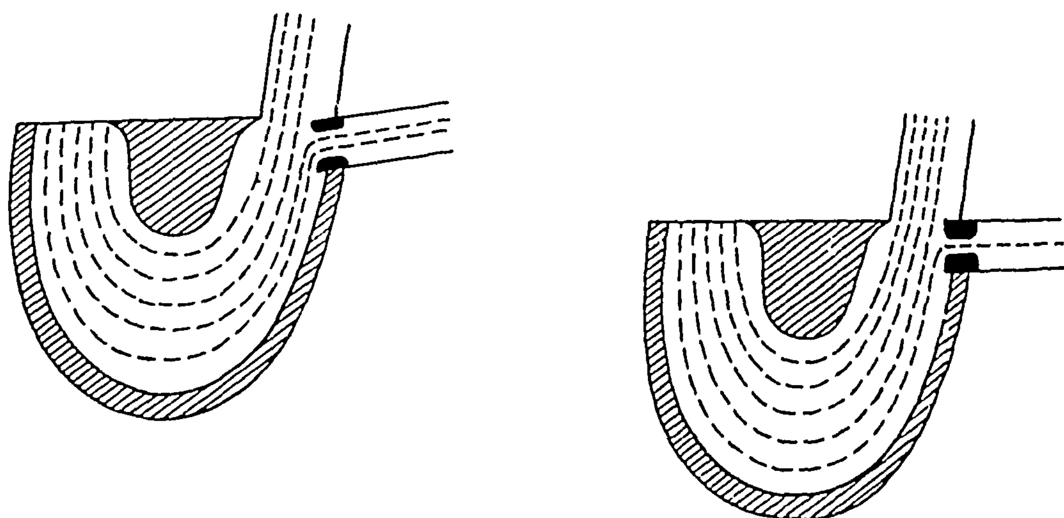
Walmesley (1929), in *Quain s Anatomy*, states that the developing myocardium reaches the transition of the bulbus into the truncus and then disappears in the distal part of the bulbus even before it is differentiated into recognisable muscle tissue. This retrogression of the bulbus myocardium is a significant feature of all vertebrate hearts, it is probably to be related to the replacement of a muscular sphincter control of the exit tube from the heart by a purely mechanically valvular control. This conclusion that the control of the pulmonary outflow is a purely mechanical function of the valve cusps and is not supported by muscular action in the mammalian heart is almost certainly incorrect; it is certainly incomplete. There is much to support Keith s view of the probable function which the muscle of the infundibulum subserves

If Keith s view is correct, the infundibulum should not contract as a whole to expel its contents but a wave of contraction should pass along it; almost a peristaltic wave. I have watched the action of the infundibulum on many hearts, normal and abnormal, exposed at operation, under favourable conditions it is possible to see quite clearly a progressive wave of contraction and relaxation passing up the infundibulum. The conditions are, however, not very favourable for recognising this and even less favourable for demonstrating it. I was, therefore, eager to study the rapid cinematographic studies of the heart made by Prinzmetal (1952) in which the heart contractions are photographed at up to 1,000 frames a second and are projected at the standard rate of 16 frames a second, thus permitting detailed leisurely study and demonstration of cardiac movements that are otherwise impossible to analyse. Prinzmetal s studies have been wholly directed to the atria, both in normal function and in arrhythmias In his films, however the whole heart can be seen and in certain places it is possible to observe quite clearly the action of the infundibular portion of the right ventricle up which a wave of contraction passes, preceded by a wave of distension or relaxation. It is unfortunately not possible to reproduce in still pictures what can be seen in the motion picture.

If it is true that the musculature of the infundibulum exercises this controlling action on the output of the right ventricle and the stability of the pulmonary valve mechanism, it follows that in addition to actual waves of muscular contraction and relaxation playing a part, muscle tonus must also be important. It is possible to make observations in pathological clinical states which tend to support this supposition that varying tonus in the right ventricular outflow tract is of functional importance.

The infundibulum in pulmonary stenosis

A characteristic clinical feature of Fallot's tetralogy is variation in the degree of cyanosis and disability, often from day to day, depending on environmental conditions, and also the occurrence of attacks of deep cyanosis and unconsciousness.



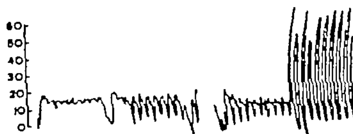
73 Diagrams to show the effect of variations in form of the pulmonary outflow tract, as the tonus increases—diagram on the right—less blood goes to the lungs and more is shunted into the aorta

These attacks are of ill-omen and may be fatal. I suggest that there is a simple explanation for their occurrence, namely increase in tonus of the infundibular musculature Thus, if the tonus of the already diminutive outflow tract increases, the narrowed lumen becomes smaller as if by sphincteric action, less blood passes to the lungs and at once the right-to-left shunt increases and more venous blood enters the aorta (Fig 73) The cyanosis is, therefore, worsened in two ways and it is not surprising that unconsciousness occurs When the stenosis is initially severe, even a small increase in tonus will be serious and it is just in those patients with severe stenosis that severe, frequent and perhaps fatal cyanotic attacks occur Presumably the tonus must relax as cyanosis becomes profound and as the coronary blood-flow becomes increasingly inadequate, general cardiac tone would inevitably fail as the coronary flow became poorer.

The milder variations in cyanosis depending on climatic and other conditions may also be due to variations in tonus of the infundibular muscle

We have been able to obtain actual direct evidence of variation in tonus of the infundibular musculature, this has emerged from catheter studies made before operation and at the time of operation Thus the tracings shown in Figure 74 can be explained in no other way In the upper tracing, made at cardiac catheterisation, a diagnosis of infundibular stenosis was made, it can be seen that there is only one change in the systolic pressure although the drop in diastolic pressure indicates clearly when the catheter passes from the pulmonary artery through the valve to enter the infundibulum. The lower tracing was made at operation and a double pressure change is shown, one at the valve level and one at the infundibulum Combined valvotomy and infundibular resection were needed The only satisfactory explanation of this is that the tonus of the infundibular muscle was greater at catheterisation

Rodbard and Shaffer (1955) have reported interesting and corroborative observations made at cardiac catheterisation on cases of Fallot's tetralogy. They





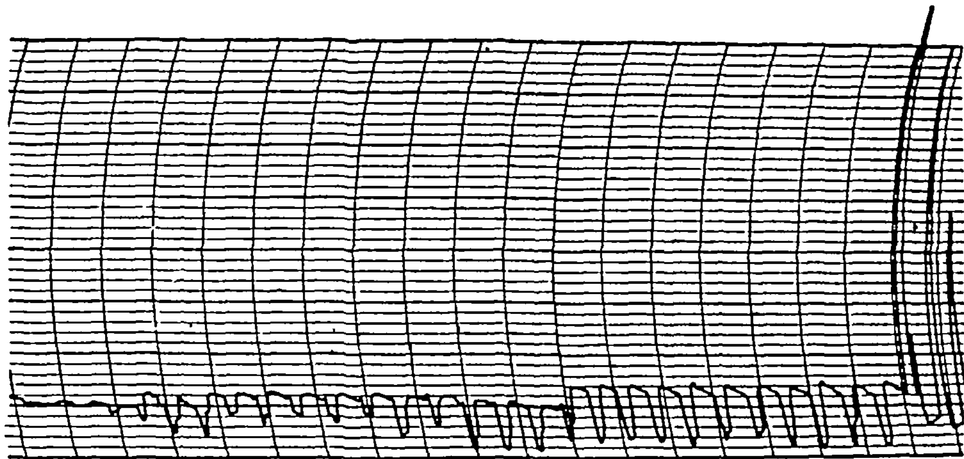
74. The top tracing taken at cardiac catheterisation shows a single stenosis—infundibular The lower tracing was taken at operation and two stenoses valvar and infundibular are shown. The only explanation is that at cardiac catheterisation the infundibular muscle was in greater tonic contraction.

observed that in 10 out of 13 cases the pressure in the right ventricle and in the infundibular chamber began to rise together for a part of the ejection phase but the infundibular pressure then began to fall, even though the right ventricular pressure continued to rise. These findings suggest that the infundibular stenosis becomes more severe during ventricular systole and Rodbard and Shaffer comment, 'Phylogenetic and electrocardiographic evidence suggests that the infundibular portion is one of the last portions of the right ventricle to become activated during systole'

They also comment that in the presence of a ventricular septal defect this dynamic sequence may well be a factor in the production of the frequently observed bidirectional shunts In early systole, shunt flow would tend to be from left-to-right. Then contraction of the infundibulum increases the resistance to pulmonary flow decreasing the shunt or even reversing it.

Certainly in Fallot s tetralogy it is likely that the degree of pulmonary stenosis increases during the later phase of ventricular systole, causing the right to-left shunt to become progressively more severe.

Figure 75 illustrates a possible trap that exists in the catheter diagnosis of infundibular stenosis Thus a withdrawal tracing seems to show the pulmonary artery pressure, then the diastolic pressure falls suggesting that the infundibulum is being traversed before the final major change to the high ventricular pressure. A diagnosis of infundibular stenosis was made. At operation a simple valve stenosis was found there was no infundibular stenosis The explanation is that the first part of the tracing shows the pressure in the right pulmonary artery;



75 Mistaken catheter diagnosis of infundibular stenosis The apparent infundibular tracing is from the pulmonary trunk in which the low diastolic pressure is due to a Venturi effect

the next part with a low diastolic pressure was actually taken from the main pulmonary artery and was due to a negative pressure in the artery and does not represent an infundibular chamber, it can be seen that it is lower than the true right ventricular diastolic pressure Dr L Brotmacher has pointed out to me that when the electromanometric tracing is studied with a synchronised electrocardiogram it can be observed that the low diastolic pressure in the pulmonary artery corresponds with the upstroke of ventricular systole, the explanation being that it is a Venturi effect due to the rapid flow of blood through the narrow orifice

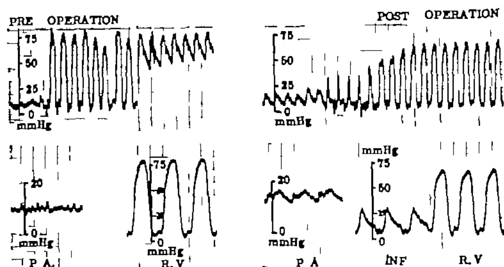
Elmslie-Smith *et al.* (1956) have also made similar observations with simultaneous reading of the intracardiac electrogram and pressure pulse in patients with pulmonary stenosis which they have found helpful in locating the level of the stenosis

Secondary infundibular stenosis after pulmonary valvotomy

Somewhat allied to the question of varying tonus in the infundibulum is the phenomenon of the development of a secondary infundibular stenosis after pulmonary valvotomy in cases of valvar stenosis with a normal aortic root, was fully described and discussed in Chapter VI

That the same development of a secondary infundibular stenosis can in Fallot's tetralogy after a successful valvotomy is shown by the observ made in Figure 76

Experiences in punch resection of infundibular ... of Fa
tetralogy also provide another indication of the pro ... e a
of the infundibular muscle Thus my ordinary pra ... sten
portion with a circular punch On occasions, ho ... type
punch resection has appeared to be inadequate I ... ich'
remove a segment of the crista supraventricularis ... e m
be immediately followed by a great increase in flow ... in t



76 Electromanometric tracings before and after pulmonary valvotomy in a case of Fallot's tetralogy to show the development of secondary infundibular stenosis just as may be seen in cases of pulmonary stenosis with intact ventricular septum. That a true Fallot's tetralogy was present is shown in the pre-operative tracing in which the catheter has entered the aorta.

pulmonary artery in fact the pressure and flow may be excessive This cannot be entirely explained by actual removal of the infundibular stenosis, it is more likely to be due to division of the sphincteric muscular mechanism and therefore complete failure of its controlling action on the outflow tract.

Discussion

It is submitted that enough evidence has been put forward to substantiate the hypothesis that a controlling mechanism exists in the musculature of the right ventricular outflow tract. When Keith originally put forward this view he recommended it to clinicians for their consideration and for application to such problems as might appear pertinent. He himself suggested only that it might be responsible for some functional systolic pulmonary bruits and lamented failure of clinicians to pay any attention at all to the facts put before them by the anatomist and morphologist (Keith, 1904a)

Basically from the point of view of pure science alone, it is important to recognise the presence of such an important function as a mechanism for influencing and controlling the pulmonary outflow from the right ventricle. There are other applications than the single one mentioned by Keith

For instance it is noteworthy that the pulmonary valve regurgitation occurs rapidly when the pressure in the pulmonary artery rises only slightly This may well be an important mechanism causing or aggravating cardiac distress following cardiac operations, any mechanical interference causing pressure on or distortion of the pulmonary artery could be very undesirable and could throw a great extra strain on the right ventricle. In the commoner causes of raised pulmonary artery

pressure there is time for compensation to occur, the secondary muscular hypertrophy of the right ventricle simultaneously improves the force of the muscular support of the valve mechanism and so cancels the tendency to regurgitation Eventually, however, as is so well known, decompensation occurs and pulmonary regurgitation develops In perhaps its commonest form this is seen in the pulmonary hypertension due to mitral stenosis and the eventual onset of pulmonary regurgitation as shown by the Graham Steell bruit, or in the pulmonary hypertension secondary to a ventricular septal defect

Acute strain may be caused in association with direct operations for pulmonary stenosis For instance, after infundibular resection in cases of Fallot's tetralogy an incision has been made into the outflow tract of the right ventricle and part of the wall has been cut away, it is not surprising that some impairment of muscle tone follows and temporary pulmonary regurgitation can occur. This may be recognised by the diastolic murmur for some days after operation, the murmur gradually fading as tone recovers and compensation occurs Although pericardial friction may mimic the murmur of valve regurgitation there can be no doubt that true temporary regurgitation occurs It is more likely to occur in a significant form if there is a thin-walled infundibular chamber distal to the stenosis and if this is suddenly exposed to a high pressure from too generous relief of the stenosis If this is so the thin muscle in the infundibulum at the valve level may stretch considerably, thus causing quite severe and prolonged pulmonary regurgitation I have certainly seen this in one case in which the pulmonary artery systolic pressure rose to 40 mm Hg, this is the only severe example I have seen in cases of infundibular resection Although it is put forward as an objection to the direct operations for Fallot's tetralogy it is not a serious one because of its rarity, provided the stenosis is not relieved so much that the thin infundibulum receives the full ventricular pressure

I have recently seen a quiet pulmonary diastolic murmur develop during pregnancy in a young woman who had undergone successful open resection of a pure infundibular stenosis one year before Presumably this was due to the increased blood-flow of pregnancy in the presence of the rather thin-walled infundibular chamber which had been present distal to the previous stenosis The right ventricular pressure, as such, had been reduced to normal

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